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LANCASTER DOCTORATE IN CLINICAL PSYCHOLOGY

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Doctoral Thesis

**Psychological distress in the context of Huntington's disease**

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## Word Count

Section	Text	Tables, Figures, Reference Lists, Appendices	Total
Thesis Abstract	283	0	283
Literature Review	7966	6587	14,553
Research Paper	7895	5516	13,411
Critical Appraisal	3368	429	3797
Ethics Proposal	4752	6828	11,580
Total	24,264	19,360	43,624

## Thesis Abstract

This thesis is comprised of a scoping literature review, a research paper and critical appraisal which focus on psychological distress and psychological therapy in the context of Huntington's disease (HD). The literature review is a scoping review of 29 papers looking at different aspects of irritability in the context of HD. The review examines the validity of irritability as a meaningful construct in HD. Clinical and theoretical implications as well as suggestions for further research are also discussed.

The research paper investigates understandings of psychological distress in HD from the perspective of people with HD as well as seeking to understand people's perspectives of psychological therapy. Semi-structured interviews were conducted with nine participants, prior to commencing a trial of mindfulness-based cognitive therapy (MBCT) and the data subsequently analysed using interpretative phenomenological analysis (IPA). Three themes emerged from the data: (1) Attributing psychological distress to HD: *"you're blaming everything on that now"*; (2) Attribution across time: *"in the past you'd just get on with it"*; (3) Therapy instils hope and fight: *"a light at the end of the tunnel"*. The results are then discussed in terms of implications for the potential for psychological services to be available to people with HD alongside the need for further research into the acceptability of psychological approaches in the context of HD. The research paper highlights a predominant biological understanding of psychological distress with a more implicit psychological understanding presented, and a hope for psychological therapy to enable people to regain control over their experience.

Finally, the critical appraisal reflects on some of the process issues encountered during the research including the impact of attending the MBCT group on the data analysis and barriers to recruitment.

## **Declaration**

This thesis presents work undertaken in partial completion of the Doctorate in Clinical Psychology at Lancaster University from May 2015 to May 2016. The work in this thesis is that author's own, except where due reference is made, and has not been submitted for any other academic award.

Name: Rachael Theed

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## **Acknowledgements**

Firstly, I would like to thank everyone who participated in this study for giving their time to be interviewed and making the research possible. It was a pleasure to work with them and hear about their experiences. I would also like to thank my supervisors, Dr Jane Simpson and Dr Fiona Eccles for their invaluable guidance and support throughout the research.

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**Section 1: Literature Review**

**Validity of irritability in Huntington's disease: A scoping review**

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**Abstract**

**Purpose:** To scope the literature concerning irritability in Huntington's disease to determine whether or not irritability is a valid and meaningful construct within this population.

**Method:** Scoping literature review.

**Results:** The review highlighted several aspects of irritability in HD which influence the validity of irritability as an independent construct within HD. Various measures are used to assess irritability yet there remains no gold standard and consequently irritability is assessed inconsistently. Irritability does not seem to reflect the HD disease process and appears to be strongly associated with other psychological constructs including depression, anxiety and apathy.

**Conclusions:** Irritability as a construct continues to lack clarity and is used and measured inconsistently. Consequently, further research is required in order to determine the extent to which irritability is a valid construct within the context of HD.

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### Validity of irritability in Huntington's disease: A scoping review

It has been suggested that irritability is commonly experienced by people with neurological conditions such as Parkinson's disease (PD; Aarsland et al., 1999), dementia (Burns, Folstein, Brandt & Folstein, 1990) and progressive supranuclear palsy (PSP; Gerstenecker, Duff, Mast, Litvan & ENGINE-PSP Study Group, 2013). However, it is perhaps most notably discussed in people with Huntington's disease (HD; Wagle, Wagle, Markova & Berrios, 2000) where it is often reported as a 'neuropsychiatric' symptom of the HD process. Moreover, many studies have reported high rates of irritability in HD (Craufurd, Thompson & Snowden, 2001; van Duijn, Kingma & van der Mast, 2007). However, it has been argued the concept as it is currently lacks psychological rigour and, as such, research and measures could be potentially measuring different concepts, for example anger and aggression (Craig, Heitanen, Markova & Berrios, 2008).

### **Introduction to Huntington's disease**

HD is an inherited neurodegenerative disease, characterised by a triad of progressive difficulties in motor, cognitive and behavioural domains (Craufurd et al., 2001). A formal diagnosis of HD is made when motor symptoms become apparent (Tabrizi et al., 2009). Age of onset (when motor symptoms start to develop) usually occurs around the age of 40 with the disease subsequently progressing over 15-20 years (Novak & Tabrizi, 2010). However, psychological difficulties are frequently experienced by people with HD prior to this onset of motor symptoms (Duff et al., 2007; Roos, 2014). Difficulties associated with HD vary across disease stages, with psychological difficulties such as irritability, depression and anxiety argued to form the three core difficulties experienced by people with HD (Kloppel et al., 2010).

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### **Conceptualising Irritability**

Irritability, in general, has been characterised as a readiness to react excessively to negative stimuli often having both an affective component, anger and behavioural component, aggression (Buss & Durkee, 1957; Caprara et al., 1985), however it is poorly defined. Snaith, Constantopoulos, Jardine and McGuffin (1978) defined irritability, in general, as a psychological state characterised by poorly controlled anger resulting in aggression, impatience and intolerance. However, in an attempt to provide a formal definition, Snaith and Taylor (1985) later proposed a definition of irritability as a “feeling state characterised by reduced control over temper which usually results in irascible verbal or behavioural outbursts, although the mood may be present without observed manifestation” (p.128). This seems to be inconsistent with psychological theory which differentiates between an emotion and a mood, seeing them as closely related yet distinct phenomena (Beedie, Terry & Lane, 2005). They further noted “it may be experienced as brief episodes...or it may be prolonged and generalised...irritability is always unpleasant for the individual and overt manifestation lacks the cathartic effect of justified outbursts of anger” (Beedie et al., 2005, p.128).

More recently, Craig et al. (2008) conceptualised irritability as a mood state, differentiating this from emotions such as anger which tend to be more reactive compared with a mood which they understood to be more prolonged. Conversely, irritability has also been conceptualised as a stable personality trait (Buss & Durkee, 1957). For example, early German psychopathologists referred to changes in behaviour, such as irritability, as part of personality change (Craufurd & Snowden, 2014). Indeed, it is evident that there are opposing views as to whether irritability should be conceptualised as a state or trait (Burns et al., 1990), or if it has elements of both.

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Indeed, the debate remains regarding whether irritability should be understood as a mood disorder independent of others such as anxiety and depression (Mangelli et al., 2006). In a study investigating irritability in physical illness including cardiovascular, cancer and endocrine illnesses, Mangelli et al. (2006) found irritability and depression to be two distinct phenomena despite some overlap. They argued this provided support for the earlier findings of Snaith and Taylor (1985) that irritability was an independent mood as opposed to one representative of anxiety or depression.

Irritability in HD, specifically, has been conceptualised using the definitions applied to the general population. However, the occurrence of irritability in HD can be more difficult to determine due to the brain changes associated with HD, potential differences in understandings of irritability and the lack of reliable methods of assessment (Craufurd & Snowden, 2014).

### **Causes of Irritability in HD**

In addition to the variety of definitions of irritability and measures used to assess it, various explanations have been put forward regarding the cause of irritability in HD (Craufurd & Snowden, 2014). It is commonly understood that irritability is the result of the biological progressive neurodegenerative nature of HD. Indeed it has been suggested that higher levels of irritability in people with HD, compared with spouse controls in the same environment, “implicates a neurobiological, rather than psychological or reactive, basis for these behavioural signs” (Tabrizi et al., 2009, p.799). For example, it has been suggested that the degeneration in areas of the brain that control socially appropriate behaviour may result in irritability in the earlier stages of HD (Mega & Cummings, 1994). Moreover, this is consistent with wider understandings that neurodegenerative changes resulting from HD are important in the development of psychological difficulties experienced by people with HD.

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While irritability is frequently identified as a separate difficulty experienced in HD, there has also been debate that it may be secondary to other psychological difficulties such as depression (Craufurd & Snowden, 2014; van Duijn., 2010). Furthermore, some people with HD often report periods of suicidal ideation after episodes of heightened irritability (Craufurd & Snowden, 2002), indicating a potential association between irritability and suicidality. On the other hand, it has been suggested that irritability may, at least in part, be a psychological consequence of difficulties with communication and cognition (Craufurd & Snowden, 2014). Difficulty communicating would understandably lead to frustration, over time resulting in an increased level of irritability.

Although the dominant perspective is that of a biological understanding, behaviour in HD is also likely to reflect both intrinsic and reactive changes (Craufurd & Snowden, 2014). Consequently, further investigation is required of the concept of irritability in HD to understand whether the occurrence of irritability is a result of alterations in the brain, in response to living with a distressing disease or a combination of the two. Indeed, irritability is noted to be a key determinant in people with HD moving to residential care (Craig et al., 2008) and as such it seems important to understand the concept to reduce the impact it has on people with HD. Furthermore, the influence different measures have on reports of irritability should also be considered.

### **Validity of irritability**

There are several types of validity important in terms of establishing whether a construct is valid, for example construct validity, predictive validity, convergent validity and discriminant validity (Kendell, 1975). Construct validity refers to how much the concept of irritability as a symptom in HD is reflective of a true difficulty that people with HD experience (Kendell, 1975).

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As part of assessing construct validity, which could be seen as a superordinate value, a number of other types of validity can also be assessed. For example, convergent validity refers to the degree to which a construct is similar to another construct that it should be similar to. In the case of the current review, this relates to whether irritability is similar to constructs such as anger and hostility. Additionally, in terms of the measures used to assess irritability, it is important to consider whether they measure irritability in a similar way or whether they distinguish between different constructs. Conversely, if it were apparent that irritability was to diverge in meaningful ways from other constructs then it could be suggested that irritability has discriminant validity. Discriminant validity refers to whether or not a diagnosis or measure can distinguish between the construct we are interested in and other constructs (Kendell, 1975). Finally, predictive validity is the extent to which a diagnosis provides useful information regarding an individual's future (Kendell, 1975). For example, reviewing the literature and considering whether or not irritability is able to predict important aspects of HD such as quality of life and disease progression.

In order to examine the concept of irritability the current review adopts a scoping review method. A scoping review aims to provide an overview of background information pertaining to an area of inquiry which can later inform a more specifically focussed systematic review (Armstrong, Hall, Doyle & Waters, 2011). Scoping reviews are particularly useful for areas which are complex and have not been extensively reviewed (Mays, Roberts & Popay, 2001). Indeed, irritability in HD has received little attention and is yet to be fully understood. As such, this scoping review aims to provide a broad overview of the existing literature on irritability in HD to investigate whether irritability could be argued to be a valid construct within this population. Consequently, this paper will review the key findings from the research, how irritability is associated with other psychological difficulties, potential treatment options for people with HD and the potential aetiology of HD. Finally, it

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will conclude whether or not irritability is a valid construct and clinically meaningful for people with HD.

### **Method**

This scoping review followed the stages outlined by Arksey and O'Malley (2005). A research question was developed and relevant studies identified. Studies appropriate for inclusion were selected and the relevant data charted in order to collate the necessary data. The results were then summarised and reported.

### **Searching for studies**

The papers selected for inclusion in this scoping review met the following inclusion criteria: (i) the paper was published in English language; (ii) published in a peer reviewed journal; (iii) the paper involved the investigation of irritability in HD including prevalence, associations with other variables, across disease stage, treatment options and aetiology. Additionally, papers investigating irritability in HD and other neurological conditions were excluded if they did not report findings for each separate neurological condition.

Relevant papers were identified by conducting a search in the databases Academic Search Complete (searchable years 2002-2014, 'peer reviewed' and 'English' selected), PsycINFO (searchable years 1940-2015, 'peer reviewed' and 'English' selected), CINAHL (searchable years 1999-2009, 'peer reviewed' selected), Scopus (searchable years 2006-2015, 'English' and 'Journals' selected), and Web of Science (searchable years 1990-2015). The search was conducted in November 2015 and the full-text search terms used to identify potential papers were "irritability" and "Huntington\*". This database search returned 334 papers (Academic Search Complete = 29, PsycINFO = 60, CINAHL = 3, Scopus = 179 and Web of Science = 63). Duplicates were removed, leaving 217 papers. Remaining papers

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were then reviewed for their suitability by reading the titles and abstracts. However, for those papers where suitability was unclear, the full text was read and subsequently the inclusion and exclusion criteria applied. This process revealed 29 papers suitable for inclusion in the current review. Table 1 provides a summary of these papers. Quality appraisal is not required within the process of a scoping review therefore was not conducted (Arksey & O'Malley, 2005).

(<Insert Table 1 here>)

### **Results**

Twenty-nine papers were included in the current review with a summary of the results presented in Table 2. Of the 29 papers, 10 compare irritability in people with HD to healthy controls, nine examine changes in irritability across disease stage, two compare irritability in individuals with HD with those with other neurological conditions, 12 report associations with other psychological difficulties in HD, three describe interventions and three report potential neurological pathways for irritability in HD. Some of these papers investigate more than one area pertaining to irritability in HD. In addition, the measures used to assess irritability are also discussed below.

(<Insert Table 2 here>)

### **Measures of Irritability**

Various measures have been developed to assess irritability both in non-HD and HD populations, for example, the Buss-Durkee Hostility Inventory (BDHI) and the Problem Behaviours Assessment for HD (PBA-HD). However, it remains that there is no gold standard for assessing irritability (Bouwens, van Duijn, van der Mast, Roos & Guiltay, 2015). Furthermore, the lack of a core and widely understood construct means that different

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measures potentially measure different constructs. This can result in inconsistencies in research findings based on the choice of measures as opposed to true differences based on issues unrelated to sampling error. Furthermore, irritability measures can rely on either self-report, caregiver-report, clinician-based assessment and in some cases a combination of the three.

A number of measures are used to assess irritability in HD (see Table 3). The Irritability, Depression, Anxiety Scale (IDA; Snaith et al., 1978) was initially developed to address the need for scales to assess irritability in clinical populations and has been used in studies assessing irritability in HD (Berrios et al., 2001; Berrios et al., 2002; Nimmagadda, Agrawal, Worrall-Davies, Markova & Rickards, 2011). Snaith et al. (1978) described irritability as a two-dimensional construct, which led to the formation of two subscales in the IDA: outwardly expressed irritability and inwardly expressed irritability (Snaith & Taylor, 1985). Additionally, this scale is reliant on self-report, thus assessing subjective irritability. Snaith and Taylor (1985) examined irritability in clinical populations, across four studies including people experiencing depression, anxiety, mood disorder and obsessional neurosis, which subsequently indicated that irritability should be understood as a mood state rather than a personality trait.

(<Insert Table 3 here>)

Further measures that have been used to assess irritability in HD include self-report measures such as the PBA-HD (Craufurd et al., 2001) and the behavioural section of the Unified Huntington's Disease Rating Scale (UHDRS-b) and informant report measures such as the John Hopkins Irritability Scale and the Burns Irritability Scale (BIS; Burns et al., 1990). Burns et al. (1990) argue that self-report measures are not suitable for people who go on to develop cognitive impairment, potentially suggesting that people lack insight into their

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own problems and thus require more objective measures. As such, informant report measures can be used alongside self-report measures to provide a more accurate account of a person's experience of irritability. The BIS (Burns et al., 1990) purports to allow for an objective measure of irritability to be obtained from a carer or family member, aiming to measure a change in behaviour in the context of illness. Therefore, someone who has always been irritable would be unlikely to score highly for irritability using this scale (Burns et al., 1990) due to the focus on change in irritability as opposed solely to the current level.

One of the most commonly used measures is The Unified Huntington's Disease Rating Scale (UHDRS; Huntington Study Group, 1996) which measures motor, cognitive and behavioural aspects of HD as well as functional capacity. Indeed, this scale was used in a number of studies included in the current review (Banaszkiewicz et al., 2012; Hubers et al., 2013; Reedeker et al., 2012; Rickards et al., 2011; Thompson, Snowden, Craufurd & Neary, 2002; Van Duijn et al., 2014).

Another measure frequently used to measure irritability in HD is the PBA-HD (Craufurd et al., 2001). This is a semi-structured interview used with both people with HD and close others such as family members. The scale comprises of three factors: apathy, irritability and depression, all with individual sub-scale items. Irritability items include inflexibility, preoccupations, irritability and verbal and physical aggression (Craufurd et al., 2001). Items are measured on a five-point scale to assess both the frequency and severity of behavioural difficulties in HD and multiplied to obtain an overall score (Gregory et al., 2015).

Indeed, it is evident that the way in which different measures conceptualise, and subsequently measure, irritability has differed. Although multi-item irritability measures such as the IRQ have been shown to have good reliability and assess a variation of thoughts,

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feelings and behaviours related to irritability (Holtzman, O'Connor, Barata & Stewart, 2015), shortcomings have been highlighted. Holtzman et al. (2015) note how scales attempting to measure irritability also tap into constructs such as anger and hostility. This is problematic in the assessment of irritability since irritability, unlike anger, often occurs in the absence of a direct cause and is longer in duration (Beedie et al., 2005), therefore implying a different construct. If irritability is to be understood as a mood state then it should be distinguished from anger which often has a clear antecedent (Craig et al., 2008). Although it is generally acknowledged that irritability is a construct distinct from anger and aggression, this is not currently reflected in the measures used to assess it (Holtzman et al., 2015).

In addition, if irritability is conceptualised as a “temporary psychological state” (Snaith et al., 1978, p.164) then it seems necessary that the individual is central in the rating process (Holtzman et al., 2015). Self-report measures, where possible, are one of the key methods of measuring irritability in HD. They are important in order to understand and measure the personal experience of the individual which is not always observable by family members or clinicians, since their ratings are based on observable behaviour (Bogart, 2011). However, in contrast to this, consideration should be given to the fact that with conditions such as HD which affect the brain, there is a potential for self-awareness to become reduced further along in the disease process (Kirkwood et al., 2002b). As such, there may be a limit to the validity of self-report measures.

Indeed, it may be difficult to measure irritability in HD accurately, particularly during more advanced stages whereby an individual may not be able to provide a self-report measure due to other difficulties that occur with the progression of HD such as cognitive impairment (Fisher, Sewell, Brown & Churchyard, 2014). As such, it may be at this point that family member and clinician-based measures are more appropriate, either as a stand-alone measure

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or in addition to self-report measures. However, if the notion of inward irritability, that which cannot be observed, is to be considered then this is likely to go un-measured due to the reliance on self-report measures (Snaith et al., 1978). Consequently, reported rates of irritability in HD may depend on the measures used, how irritability is defined and the stage of disease (van Duijn et al., 2013). Measures such as the PBA-HD which are conducted with the person with HD, a spouse or carer and acknowledge observations made by the interviewer (Callaghan et al., 2015), may be most appropriate. Furthermore, it may also be important to consider whether a single measure of irritability is able to provide an accurate depiction of irritability (Kloppel et al., 2010).

### **Irritability in people with HD compared with healthy controls**

A range of studies have compared irritability in people with HD with healthy controls (Berrios et al., 2001; Berrios et al., 2002; Julien et al., 2007; Kirkwood et al., 2002a; Kirkwood et al., 2002b; Kingma, van Duijn, Tinman, van der Mast & Roos, 2008; Kloppel et al., 2010; Reedeker et al., 2012; van den Stock et al., 2015; Vassos, Panas, Kladi & Vassilopoulos, 2007). Seven of the 10 studies found that irritability is significantly higher in people with HD compared with healthy controls (Berrios et al., 2001; Berrios et al., 2002; Julien et al., 2007; Kingma et al., 2008; Kirkwood et al., 2002a; Reedeker et al., 2012; Van den Stock et al., 2015). In a review of the prevalence of psychological difficulties in HD, van Duijn et al. (2007) found reported rates of irritability in people with HD to range from 38% to 73% as measured by the PBA-HD and NPI. As cut off scores are used to determine whether irritability is present, and to what extent, it is apparent there is an assumption that irritability is a symptom of HD which can be diagnosed. However, due to no standardised cut off score existing for measuring irritability, across different measures, those selected are at the discretion of the researcher.

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Additionally, irritability levels have been found to vary in accordance with report type. There was agreement between both self-report and informant-report that 28% of those with the expanded gene were considered irritable and 50% were not irritable. With regards to the remaining participants, there was disagreement between people with HD and their informants with informants reporting more irritability in people with HD compared with the individuals themselves (Reedeker et al., 2012). This could potentially be a result of people with HD having reduced insight into their experience and presentation.

In addition, Kirkwood et al. (2002a) observed an increase in irritability and clinical hostility over an average of a 3.7-year period in pre-symptomatic gene carriers compared with non-gene carriers. This demonstrated that irritability may occur prior to the occurrence of physical clinical symptoms. Similarly, Berrios et al. (2002) found that gene carriers had a significantly higher level of both inward and outward irritability than non-gene carriers measured by the SIS, which loaded onto the 'personality' factor suggesting irritability may be part of a personality change occurring in HD.

However, three out of the ten studies which compared people with HD to healthy controls failed to find a significant difference in irritability (Kirkwood et al., 2002b; Kloppel et al., 2010; Vassos et al., 2007). Kloppel et al. (2010) did not find a significant difference in irritability between pre-symptomatic gene carriers and non-gene carriers. Additionally, there was agreement between the pre-symptomatic gene carriers group and their close companions regarding their level of irritability. Due to the inclusion of informant-report in addition to self-report, it may be suggested that the potential for under-reporting and limited insight is reduced, thus reflecting a more accurate depiction of their irritability.

Similarly to Kloppel et al. (2010), Kirkwood et al. (2002b) did not find a difference in irritability between those with manifest HD, pre-symptomatic gene carriers and non-gene

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carriers as measured by the MMPI. It is noted that the MMPI may not be sensitive to changes observed in HD (Kirkwood et al., 2002b) due to it being a measure of personality traits and psychopathology used within non-HD populations. However, the use of the SIS constructed for use with clinical populations, in the study by Kloppel et al. (2010), was not able to detect differences in irritability between pre-symptomatic gene carriers and non-gene carriers. It may therefore be argued that the selected measure was not the only reason for lack of significant results. Additionally, the control group in this study were companions of those with HD and thus indirectly affected by HD. Therefore, their experience of irritability may differ to healthy controls who are not affected by HD.

Finally, Vassos et al. (2007) investigated the psychological and behavioural features which differentiate people with HD from those without HD. Within this study they did not find a significant difference in either inward or outward irritability as measured by the SIS. This is in contrast to the findings of Berrios et al. (2002) who also used the SIS as a measure of irritability. On examining the effect size for the study by Vassos et al. (2007), a small effect size of  $d = 0.20$  for inward irritability suggests an effect is potentially detectable, however an effect size of  $d = 0.06$  for outward irritability suggests there is no difference to find. Indeed, the sample included by Vassos et al. (2007) was  $n = 64$  while Berrios et al. (2002) had a sample size of  $n = 98$ . However, Vassos et al. (2007) did find that people with HD showed a significantly higher level of extroverted hostility compared with healthy controls, describing hostility as a personality dimension rather than a behavioural aspect. Similarly, Berrios et al. (2002) found that both inward and outward irritability loaded onto a personality factor for people with HD within their factor structure suggesting irritability may be part of a person's personality.

### **Irritability across disease stage**

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While gene carriers have been compared with healthy controls, it has been suggested that irritability varies across stage of disease, potentially with those closer to the onset of clinical symptoms becoming increasingly more irritable. In addition, irritability may reflect a change in people with HD for a substantial period prior to the onset of motor symptoms (Julien et al., 2007). As such, a comparison of people with HD across disease stage seems appropriate in order to understand the trajectory of irritability in HD.

Of the ten papers included in this review comparing irritability across disease stage, seven did not find a significant difference in irritability across disease stage (Bouwens et al., 2015; Craufurd et al., 2001; Julien et al., 2006; Kingma et al., 2008; Kirkwood et al., 2002b; Pflanz, Besson, Ebmeier & Simpson, 1991; van Duijn et al., 2013). Included among the more cross sectional studies were two longitudinal studies which also found no significant increase in irritability between baseline (two years after entering the study) and two-year follow-up (Bouwens et al., 2015; van Duijn et al., 2013). Bouwens et al. (2015) took measurements of irritability at two time points using the Irritability Scale (Chatterjee, Anderson, Moskoqitz, Hauser & Marder, 2005) and found of those who were irritable at baseline, i.e. two years after entering the study (33%), 70% remained irritable at follow-up two years later. Furthermore, of those who were not irritable at baseline, only 23% went on to report irritability at follow-up.

Similarly, in a study by Craufurd et al. (2001), no linear relationship emerged between irritability and disease duration. However, across a disease duration span of 1-23 years, they found that difficulties defined under the factor 'irritability', including irritability, verbal aggression, physical aggression, inflexibility and pathologic preoccupation, occurred more frequently in people with a disease duration of 6-11 years. Interestingly, they found a linear relationship between apathy and disease duration which they argued might suggest apathy

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reflects a disease process whereas irritability may not. Consistent with the finding that apathy increases with disease progression, Kingma et al. (2008) found that both early symptomatic and advanced symptomatic gene carriers did not reveal more irritability than pre-symptomatic gene carriers, however did show more apathy the further advanced in disease they were. Again, this suggests that irritability may not be an underlying process associated with the disease process of HD, rather it is in response to living with HD.

However, three papers did find a difference in irritability across disease stage (Gregory et al., 2015; Thompson et al., 2012; van Duijn et al., 2014), although their findings were not consistent. Irritability was found to be significantly higher in those with clinically diagnosed early HD when compared with people with pre-manifest HD. However, this research was not extended to those with more advanced HD (Gregory et al., 2015). Van Duijn et al. (2014) found that in people with HD who experienced moderate to severe irritability, this increased by stage of disease from 10.4% at stage 1 to 19.6% at stages 4-5. However, this increase at such an advanced stage could potentially be seen as general distress due to the impact of HD at this stage. Similarly, a longitudinal study by Thompson et al. (2012) showed an increase in irritability over time as measured by the PBA-HD. Irritability was marked as being present if participants scored a severity score of greater than or equal to 2. However, this was limited to a significant linear effect in those who entered the study at stage 1 and 2 but not in those who entered at stage 3 of HD. The progression of irritability was therefore only evident in early stage HD. On discussing these findings, Thompson et al. (2012) note that irritability was common among their sample, further describing poor temper control in 80% of participants and physical aggression in 50%. Indeed, it appears that temper and aggression, frequently measured independently of irritability are assumed, in this study, to be aspects of irritability as opposed to separate constructs.

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However, when interpreting the findings, consideration should be given to studies in which participants were taking medication to manage their irritability and the impact this may have had on reports of irritability. Participants in the study by Thompson et al. (2012) had access to psychiatric input and therefore may have been taking medication to manage their irritability. As irritability only increased during the early stages of HD, it is possible that people were prescribed medication when it started to impact on their quality of life. Consequently, if irritability is managed in the later stages then it is possible that the level of irritability may not continue to progress due to medication being accessed. Similarly, Craufurd et al. (2001) reported 35% of participants to be taking medication to manage irritability. Consequently, differences in findings across studies may be influenced by the current treatment options being accessed by people with HD included in the studies.

### **Comparing HD with other neurodegenerative conditions**

Since irritability has been reported to occur in neurological conditions other than HD, it seems appropriate to compare irritability in people with HD with people with other neurodegenerative conditions. Burns et al. (1990) compared people with HD with people with Alzheimer's Disease (AD) on irritability and apathy using an irritability/apathy scale developed for their research. They found no significant difference in irritability (58%) or apathy (48%) between the HD and AD groups. The HD group were significantly more aggressive than the AD group with their aggressive outbursts lasting longer. Additionally, in both groups, irritability, apathy and aggression appeared to be independent of each other, thus suggesting an increase in one difficulty would not predict the level of another. Interestingly, irritability correlated positively with bad temper in the HD group while there was no correlation in the AD group. Thus, while there was no significant difference between the two

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groups, people with HD demonstrated higher levels of aggression and bad temper than those with AD.

In addition, Litvan, Paulsen, Mega and Cummings (1998) compared people with HD to people with Progressive Supranuclear Palsy (PSP) using the NPI. Irritability was shown to influence the total NPI score in people with HD. Additionally, the HD group scored significantly higher on agitation, irritability and anxiety while those with PSP scored higher for apathy. In the HD group agitation was positively correlated with anxiety, irritability, disinhibition and euphoria. Similarly, irritability was associated with anxiety, disinhibition, euphoria and depression. On comparing the two groups, logistic regression analysis indicated that people with HD were more likely to exhibit hyperactive behaviour (agitation, irritability) whereas people with PSP were more likely to exhibit hypoactive behaviour (apathy). These findings support the earlier findings of Burns et al. (1990) which reported that irritability and apathy can occur independently of each other. Correspondingly, the development of more recent measures such as the PBA-HD and results of factor analyses (Craufurd et al., 2001; Rickards et al., 2011) support the notion that irritability is a distinct independent difficulty experienced by people with HD.

### **Association with other psychological difficulties**

Research into irritability has also often been investigated along with other psychological difficulties reported to be common in HD. In light of findings comparing people with HD with other neurological conditions suggesting two independent processes, it seems necessary to examine this in more detail. In order to examine the associations between irritability and other psychological difficulties, correlations were examined. Of the twelve studies comparing irritability with other psychological difficulties in HD, eight reported correlations between irritability and other psychological difficulties. Seven of these studies

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reported significantly positive correlations with other psychological difficulties including apathy (Bouwens et al., 2015; Pflanz et al., 1991), anxiety (Litvan et al., 1998; Nimmagadda et al., 2011; Paulsen, Ready, Hamilton, Mega & Cummings, 2001), depression (Litvan et al., 1998; Nimmagadda et al., 2011; van Duijn et al., 2014) and bad temper (Burns et al., 1990). Only one paper reported no correlation between irritability and cognitive impairment (Thompson et al., 2002).

In addition, one paper found irritability was significantly positively correlated with suicidal ideation at baseline (Hubers et al., 2013) however this was not maintained at four-year follow up and thus was not an independent predictor of suicidal ideation. Furthermore, Banaszekiewicz et al. (2012) found irritability was not significantly related to functional disability.

However, Bouwens et al. (2015), in a longitudinal analysis, demonstrated that an increase in irritability was associated with an increase in apathy over a two-year period, an association that was maintained after other variables had been controlled for. Subsequently it was suggested that while irritability is often linked to the outward expression of anger, irritability may also only be expressed internally (Bouwens et al., 2015), similar to the proposition by Snaith and Taylor (1985) who developed the IDA to examine both inward and outward irritability. Therefore, for some people with HD, irritability may be expressed inwardly and, as such, be experienced in a similar way to apathy. Consequently, apathy has the potential to mask irritability whereby an individual may be internally distressed by feelings of irritability, without it being expressed overtly.

Furthermore, three studies found associations between irritability and anxiety. Both Litvan et al. (1998) and Paulsen et al. (2001) found irritability to be significantly positively correlated with anxiety,  $r = 0.88$  and  $r = 0.43$  respectively, as measured by the NPI.

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Similarly, Nimmagadda et al. (2011) also found that participants' inward and outward irritability scores were both significantly positively associated with both their state and trait anxiety as measured by the IDA and STAI. These correlations suggested the more anxiety a person felt, the more irritable they felt. While the causal relationship cannot be determined here, it may be suggested that irritability can occur as a result of or in response to feelings of anxiety. Therefore, people with HD who have higher levels of anxiety may be more prone to become irritable. Alternatively, a high correlation such as that found by Litvan et al. (1998) may evidence that anxiety and irritability are the same construct.

In addition to apathy and anxiety, there were also associations between irritability and depression. Irritability was found to be positively correlated with depression in the study by Litvan et al. (1998) using the NPI. Additionally, Nimmagadda et al. (2011) also found irritability (both IDA-inward and IDA-outward) to be significantly positively associated with depression as measured subjectively by the IDA-D and objectively by the MADRS. However, these correlations did not persist when irritability was informant reported as measured by the BIS. Nimmagadda et al. (2011) did however note that this difference could be due to informants not recognising irritability in people with HD struggling with depression. This idea would be supported by the finding that the IDA-inward irritability score showed a stronger correlation with the depression score on the MADRS, suggesting people with depression in HD may internalise irritability and thus hide it from those around them. Interestingly, evidence suggests that a history of depression (van Duijn et al., 2014) and bad temper (Burns et al., 1990) may increase the likelihood of people with HD experiencing irritability.

### **Treatment options for irritability in HD**

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Additionally, it is also important to examine whether irritability can be improved in people with HD. Three papers looked at the treatment options for irritability in HD (Bouwens et al., 2015; Groves et al., 2011; van Duijn, 2010). Groves et al. (2011) examined the treatment of irritability in HD using an HD irritability survey developed for their research. Indeed, the evidence should be understood in the context in which it was gathered, i.e. by expert opinion rather than through a randomised control trial (RCT), and is therefore a lower level of evidence. However, their survey revealed the use of a variety of pharmacological treatments used to reduce irritability without any general consensus, particularly with regards to treatment duration. However, there was some consensus provided by expert clinicians with selective serotonin reuptake inhibitors (SSRIs) and antipsychotics (APDs) being the preferred medication. Additionally, when considering that people with HD may also experience other psychological difficulties, as discussed above, there were differences in the selection of medication based on comorbidity. SSRIs were preferred when irritability occurred with comorbid depression and anxiety whereas APDs were often used when irritability occurred alongside aggression and impulsivity (Groves et al., 2011).

As such it may be suggested that SSRIs are used more when irritability co-occurs with depression and APDs when irritability co-occurs with aggression. Indeed, given the difference in usage by clinicians, it is possible that the medication used may actually be having an effect on the comorbid psychological difficulty i.e. depression, as opposed to irritability. Considering the correlations with other psychological difficulties it is possible that this is the case and as such the treatment of these co-occurring difficulties is more effective than aiming to manage the irritability. Consequently, it could be suggested that irritability occurs as part of these other difficulties, i.e. depression and anxiety, and as such does not represent a valid individual 'symptom' of HD.

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Interestingly, Bouwens et al. (2015), in a longitudinal study, found that the use of APDs was associated with an increase in irritability over a two-year period. However, it cannot be ruled out that APDs were initially used due to the presenting level of irritability as opposed to increasing it. Indeed, this would concur with the evidence suggesting APDs are the preferred choice of medication when anxiety co-occurs alongside aggression (Groves et al., 2011).

### **Suggested neurological pathways for irritability in HD**

Research has also examined the potential neurological pathways due to the little that is known about the potential brain changes associated with the psychological aspects of HD. Three studies included in this review investigated the underlying brain changes occurring in HD (Gregory et al., 2015; Kloppel et al., 2010; van den Stock et al., 2015).

Van den Stock et al. (2015) found evidence of striatal atrophy and increased irritability in the gene positive group compared to healthy controls. They looked at the association between clinical irritability and experience of anger by correlating irritability scores on the PBA-HD with functional MRI (fMRI) activation in people who were gene positive, however not showing any motor symptoms. They found a significant positive correlation between irritability and pulvinar activation concluding that the thalamic pulvinar plays a key role in irritability in HD. Additionally, anger experience was associated with hyper-activation of the emotion experience neurocircuitry. Therefore, it is possible that the areas of the brain assumed to be activated as a result of experiencing irritability may instead be activated by the experience of anger.

Kloppel et al. (2010) found higher levels of reported irritation were associated with stronger activation of the amygdala in controls compared with pre-symptomatic gene carriers for whom correlations were absent. They argue that inappropriate responses of the amygdala

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make pre-symptomatic gene carriers increasingly prone to psychological difficulties such as irritability. Additionally, the involvement of the amygdala has also been found in research investigating aggression, highlighting a potential link for the experience of negative emotions such as irritability, anger and frustration more generally as opposed to being specific to irritability.

Furthermore, comparing people with early HD with people with pre-manifest HD, Gregory et al., (2015) found a significant negative correlation between irritability, as measured by the PBA-HD, and fractional anisotropy across the whole brain with a decrease in white matter microstructure. These findings were reversed in those closer to onset with results being maintained following controlling for medication use. Additionally, they suggested that due to the dominant involvement of the posterior tracts and left hemisphere it is possible that the increase in irritability could be a result of cognitive overload. Consequently, the evidence regarding the potential neural pathways seems unclear and potentially confounded by other psychological and cognitive aspects.

### **Discussion**

Following the review of research looking at irritability in HD, the validity of irritability as a symptom in HD can now be assessed in terms of whether this is supported by the research.

Considering that there is no gold standard for measuring irritability, cut off scores used across research often vary and, as such, remain arbitrary (Reedeker et al., 2012). For example, three studies using the irritability scale (Chatterjee et al., 2005; Kloppel et al., 2010; Reedeker et al., 2012) used varying cut off scores of >15 and >14. Although the variation in these is not great, there remains the potential for different results to be obtained. While efforts have been made to reduce the impact of this on results, it seems that if irritability is to

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be considered a symptom of HD, there should be standardised measures and scores specific for people with HD. However, difficulties with agreement regarding standardised measures and clinical cut off scores seems to be perpetuated by the, still, lack of agreed definition.

Additionally, as a result of this lack of agreed definition, it has been acknowledged that individual participants may have different understandings of irritability (Kloppel et al., 2010). Therefore, people's experience and understanding of what irritability comprises is likely to differ as people attribute different behaviours to irritability. For example, some people with HD may understand anger and aggression as a consequence of irritability whereas others may not.

Indeed, it seems apparent that it is difficult to determine whether irritability is a separate construct from those such as anger, aggression and agitation. For example, Paulsen et al. (2001) found a high correlation between irritability and agitation ( $r = 0.81$ ) suggesting the same construct was being measured and as such irritability may not, as it is currently, be a valid independent symptom of HD. This difficulty in discriminating between irritability and anger does not seem surprising when measurement is taken into account. For example, the NPI was used in a number of studies in the current review (Litvan et al., 1998; Paulsen et al., 2001), in which the item for irritability is 'does the patient have sudden flashes of anger' (Cummings et al., 1994). As such, in the study by Paulsen et al. (2001) a positive correlation between agitation and irritability may be expected as a result of the measure used to assess irritability. Siemer's (2009) dispositional theory of moods assumes that moods dispose people to appraise events/situations in an emotionally congruent manner. It may therefore be suggested that irritability may predispose an individual to become angry or make angry appraisals, consistent with how they are currently feeling.

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Additionally, irritability has been shown to positively correlate with anxiety (Litvan et al., 1998; Paulsen et al., 2001) and depression (Litvan et al., 1998; Nimmagadda et al., 2011). The findings indicate the potential for irritability to result from feelings of anxiety and depression or vice versa as opposed to it being an independent construct. Certainly it has been suggested, in irritability research in young people, that higher levels of irritability predict aggression, anxiety and depression in early adulthood (Leibenluft & Stoddard, 2013).

Interestingly, a factor analysis showed irritability to be an independent factor (Craufurd et al., 2001). However, aggression was located within this factor which may suggest these two constructs are not independent and that aggression occurs as part of irritability, potentially as an external expression. Furthermore, irritability has been noted to be “viewed as a decreased threshold for experiencing frustration” (Deveney et al., 2013, p.1187). As irritability is often elicited through tasks which induce frustration, it is possible that irritability is the expression of multiple frustrations which are likely to differ between people. Subsequently, irritability may be the result of people struggling to regulate their emotions and behavioural responses, whereby if frustrations become too much, anger and aggression follows.

Additionally, from the research reviewed here, the concept of irritability does not seem to have predictive validity. Of the ten papers that investigated irritability across disease stage, seven did not find a difference suggesting irritability is not part of the disease process. Similarly, in those that did find a difference across disease stage, this was only found for the earlier disease stages. Therefore, irritability did not follow the course of degeneration, while difficulties such as apathy did (Kingma et al., 2008). Consequently, irritability may not be a valid predictor of a person's experience of HD or the impact HD has on a person.

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Indeed, this review has highlighted the variation in assessment tools and clinical cut offs used to assess irritability in HD. As such, it may be suggested that irritability is unlikely to be consistently measured. Considering the differences used in practice and research, where one person may be assessed as being 'irritable' by one measure, they may not be by another. Consequently, it seems that irritability needs clear definition. Considering the experience of irritability may differ in the context of HD, it is possible that irritability needs to be understood and defined specific to this clinical population.

Additionally, assessing psychological difficulties in HD may be difficult as a result of the co-occurrence of physical, motor and cognitive difficulties (van Duijn et al., 2014). Consequently, consideration needs to be given to confounding factors which may contribute to a person's experience of irritability to determine what the most appropriate form of support may be. For example, treating irritability in HD in the same way as in the general population or different clinical populations may not be effective if confounding factors, potentially impacting on a person's level of irritability, are ignored.

### **Limitations and future research**

This review, however, does have limitations. Firstly, the review only included articles that were published in English. Secondly, due to the broad nature of a scoping review, and the lack of a specific question to be answered, a general overview of the literature regarding irritability in HD is presented. Consequently, more specific systematic reviews may be required in order to understand the various aspects of irritability in HD in more depth.

Future research needs to consider how irritability is understood in the context of HD. This may include further investigation into the neural pathways and circuitry associated with irritability and considering whether areas are, in fact, central to irritability or other potentially associated constructs such as anger. Furthermore, consensus should be sought regarding the

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measures used to assess irritability in HD, considering whether different measures do assess the same construct or variations of it.

### **Conclusions**

Considering the available literature, there is currently no one satisfactory definition of irritability within the context of HD. Indeed, considering the correlates of irritability, including depression, apathy and anxiety, it may be suggested that these may provide more meaningful information about a person's experience. Additionally, current treatment options, again appear designed to treat the comorbid psychological difficulties people experience rather than specifically targeting irritability. Furthermore, with regards to understanding the aetiology of irritability in HD, the research remains unclear both in terms of the biological nature and aetiology of irritability and the associations with other psychological difficulties that co-occur. Irritability may have cognitions associated with how a person feels when irritable which may subsequently lead to the overt expression of irritability as anger. Therefore, measures need to capture the associated behavioural, cognitive and affective dimensions (Eckhardt, Norlander & Deffenbacher, 2004).

Indeed, the evidence presented makes it difficult to conclude whether irritability in HD is a valid concept, with conflicting results being found. Certainly, some research has shown irritability to have convergent validity (Litvan et al., 1998; Paulsen et al., 2001) while other research has indicated that irritability discriminates from other constructs (Craufurd et al., 2001; Rickards et al., 2011). Consequently, further research is required in order to fully understand the impact irritability has on quality of life in people with HD to conclude that it is a clinically meaningful symptom.

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## References

- Aarsland, D., Larsen, J.P., Lim, N.G., Janvin, C., Karlsen, K., Tandberg, E., & Cummings, J.L. (1999). Range of neuropsychiatric disturbances in patients with Parkinson's disease. *Journal of Neurology, Neurosurgery and Psychiatry*, 67 (4), 492-496.
- Arksey, H., & O'Malley, L. (2005). Scoping studies: Towards a methodological framework. *International Journal of Social Research Methodology: Theory and Practice*, 8 (1), 19-32. doi: 10.1080/1364557032000119616.
- Armstrong, R., Hall, B.J., Doyle, J., & Waters, E. (2011). 'Scoping the scope' of a Cochrane review. *Journal of Public Health*, 33 (1), 14-150. doi: 10.1093/pubmed/fdr015.
- \*Banaszkiewicz, K., Sitek, E.J., Rudzinska, M., Soltan, W., Slawek, J., & Szczudlik, A. (2012). Huntington's disease from the patient, caregiver and physician's perspective: Three sides of the same coin? *Journal of Neural Transmission*, 119 (11), 1361-1365. doi: 10.1007/s00702-012-0787-x.
- Beedie, C.J., Terry, P.C., & Lane, A.M. (2005). Distinctions between emotion and mood. *Cognition and Emotion*, 19, 847-878. doi: 10.1080/02699930541000057.
- \*Berrios, G.E., Wagle, A.C., Markova, I.S., Wagle, S.A., Ho, L.W., Rubinsztein, D.C., ... Hodges, J.R. (2001). Psychiatric symptoms and CAG repeats in neurologically asymptomatic Huntington's disease gene carriers. *Psychiatry Research*, 102, 217-225. doi: 10.1016/S0165-1781(01)00257-8.
- \*Berrios, G.E., Wagle, A.C., Markova, I.S., Wagle, S.A., Rosser, A., & Hodges, J.R. (2002). Psychiatric symptoms in neurologically asymptomatic Huntington's disease gene carriers: A comparison with gene negative at risk subjects. *Acta Psychiatrica Scandinavica*, 105, 224-230. doi: 10.1034/j.1600-0447.2002.0o456.x.

## IRRITABILITY IN HUNTINGTON'S DISEASE

Bogart, K. R. (2011). Is apathy a valid and meaningful symptom or syndrome in Parkinson's disease? A critical review. *Health Psychology, 30* (4), 386. doi: 10.1037/a0022851.

\*Bouwens, J.A., van Duijn, E., van der Mast, R.C., Roos, R.A.C. & Giltay, E.J. (2015). Irritability in a prospective cohort of Huntington's disease mutation carriers. *Journal of Neuropsychiatry and Clinical Neuroscience, 27*, 206-212. doi: 10.1176/appi.neuropsych.14030051.

\*Burns, A., Folstein, S., Brandt, J., & Folstein, M. (1990). Clinical assessment of irritability, aggression and apathy in Huntington and Alzheimer disease. *Journal of Nervous and Mental disease, 178* (1), 20-26.

Buss, A.H., & Durkee, A. (1957). An inventory for assessing different kinds of hostility. *Journal of Consulting Psychology, 21*, 343-349. doi: 10.1037/h0046900.

Callaghan, J., Stopford, C., Arran, N., Boisse, M.F., Coleman, A., Dar Santon, R., ... Craufurd, D. (2015). Reliability and factor structure of the short Problem Behaviors Assessment for Huntington's Disease (PBA-s) in the TRACK-HD and REGISTRY studies. *Journal of Neuropsychiatry and Clinical Neuroscience, 27*, 59-64. doi: 10.1176/appi.neuropsych.13070169.

Caprara, V.G., Cinnani, V., D'Imperio, G., Passerini, S., Renzi, P., & Travaglia, G. (1985). Indicators of impulsive aggression: Present status of research on irritability and emotional susceptibility scales. *Personality and Individual Differences, 6* (6), 665-674.

\*Chatterjee, A., Anderson, K.E., Moskoqitz, C.B., Hauser, W.A., & Marder, K.S. (2005). A comparison of self-report and caregiver assessment of depression, apathy and

## IRRITABILITY IN HUNTINGTON'S DISEASE

irritability in Huntington's disease. *The Journal of Neuropsychiatry and Clinical Neurosciences*, 17 (3), 378-383. doi: 10.1176/jnp.17.3.378.

Craig, K.J., Hietanen, H., Markova, I.S., & Berrios, G.E. (2008). The Irritability Questionnaire: A new scale for the measurement of irritability. *Psychiatry Research*, 159, 367-375. doi: 10.1016/j.psychres.2007.03.002.

\*Craufurd, D., & Snowden, J.S. (2002). Neuropsychological and neuropsychiatric aspects of Huntington's disease. *Oxford Monographs on Medical Genetics*, 45, 62-94.

Craufurd, D., & Snowden, J.S. (2014). Neuropsychiatry and Neuropsychology. In G.P. Bates, S.J. Tabrizi, & L. Jones (eds.) *Huntington's Disease* (4<sup>th</sup> Ed.). Oxford: Oxford University Press.

\*Craufurd, D., Thompson, J.C., & Snowden, J.S. (2001). Behavioral changes in Huntington's disease. *Neuropsychiatry, Neuropsychology and Behavioral Neurology*, 14 (4), 219-226.

Cummings, J.L., Mega, M., Gray, K., Rosenberg-Thompson, S., Carusi, D.A., & Gornbein, J. (1994). The Neuropsychiatric Inventory: Comprehensive assessment of psychopathology in dementia. *Neurology*, 44 (2), 2308-2314. doi: 10.1212/WNL.44.12.2308.

Deveney, C.M., Connolly, M.E., Haring, C.T., Bones, B.L., Reynolds, R.C., Kim, P., ... Leibenluft, E. (2013). Neural mechanisms of frustration in chronically irritable children. *American Journal of Psychiatry*, 170 (10), 1186-1194. doi: 10.1176/appi.ajp.2013.12070917.

Duff, K., Paulsen, J.S., Beglinger, L.J., Langbehn, D.R., Stout, J.C. & the Predict-HD Investigators of the Huntington Study Group. (2007). Psychiatric symptoms in

## IRRITABILITY IN HUNTINGTON'S DISEASE

- Huntington's disease before diagnosis: The Predict-HD study. *Journal of Biological Psychiatry*, 62 (12), 1341-1346. doi: 10.1016/j.biopsych.2006.11.034.
- Eckhardt, C., Norlander, B., & Deffenbacher, J. (2004). The assessment of anger and hostility: a critical review. *Aggression and Violent Behaviour*, 9, 17-43.
- Fisher, C. A., Sewell, K., Brown, A., & Churchyard, A. (2014). Aggression in Huntington's disease: A systematic review of rates of aggression and treatment methods. *Journal of Huntington's disease*, 3 (4), 319-332. doi: 10.3233/JHD-140127.
- Gerstenecker, A., Duff, K., Mast, B., Litvan, I., & ENIGNE-PSP Study Group (2013). Behavioural abnormalities in progressive supranuclear palsy. *Psychiatry Research*, 210 (3), 1205-1210. doi: 10.1016/j.psychres.2013.08.045. C
- \*Gregory, S., Scahill, R.I., Seunarine, K.K., Stopford, C., Zhang, H., Zhang, J., ... & Track-HD Investigators. (2015). Neuropsychiatry and white matter microstructure in Huntington's disease. *Journal of Huntington's Disease*, 4, 239-249. doi: 10.3233/JHD-150160.
- \*Groves, M., van Duijn, E., Anderson, K., Craufurd, D., Edmondson, M. C., Goodman, N., ... & Goodman, L. (2011). An international survey-based algorithm for the pharmacologic treatment of irritability in Huntington's disease. *PLoS currents*, 3. doi: 10.1371/currents.RRN1259.
- Holtzman, S., O'Connor, B.P., Barata, P.C., & Stewart, D.E. (2015). The Brief Irritability Test (BITe): A measure of irritability for use among men and women. *Assessment*, 22 (1), 101-115. doi: 10.1177/1073191114533814.
- \*Hubers, A.A.M., van Duijn, E., Roos, R.A.C., Craufurd, D., Rickards, H., Landwehrmeyer, G.B., ... & The REGISTRY investigators of the European Huntington's Disease

## IRRITABILITY IN HUNTINGTON'S DISEASE

- Network. (2013). Suicidal ideation in a European Huntington's disease population. *Journal of Affective Disorders, 151*, 248-258. doi: 10.1016/j.jad.2013.06.001.
- Huntington Study Group. (1996). Unified Huntington's Disease Rating Scale: Reliability and consistency. *Movement Disorders, 11*, 136-142.
- \*Julien, C.L., Thompson, J.C., Wild, S., Yardumian, P., Snowden, J.S., Turner, G. & Craufurd, D. (2007). Psychiatric disorders in preclinical Huntington's disease. *Journal of Neurology, Neurosurgery and Psychiatry, 78*, 939-943. doi: 10.1136/jnnp/2006.103309.
- Kendell, R.E. (1975). *The role of diagnosis in psychiatry*. Oxford: Blackwell Scientific.
- \*Kingma, E.M., van Duijn, E., Tinman, R., van der Mast, R.C., & Roos, R.A.C. (2008). Behavioural problems in Huntington's disease using the Problem Behaviours Assessment. *General Hospital Psychiatry, 30*, 155-161. doi: 10.1016/j.genhosppsy.2007.11.005.
- \*Kirkwood, S.C., Siemers, E., Viken, R., Hodes, M.E., Conneally, P.M., Christian, J.C., & Foroud, T. (2002a). Longitudinal personality changes among presymptomatic Huntington disease gene carriers. *Neuropsychiatry, Neuropsychology and Behavioral Neurology, 15* (3), 192-197. doi: 10.1097.01.WNN.0000020012.79870.51.
- \*Kirkwood, S.C., Siemers, E., Viken, R.J., Hodes, M.E., Conneally, P.M., Christian, J.C., & Foroud, T. (2002b). Evaluation of psychological symptoms among presymptomatic HD gene carriers as measured by selected MMPI scales. *Journal of Psychiatric Research, 36*, 377-382. doi: 10.1016/S0022-3956(02)00054-7.

## IRRITABILITY IN HUNTINGTON'S DISEASE

\*Klöppel, S., Stonnington, C. M., Petrovic, P., Mobbs, D., Tüscher, O., Craufurd, D., ... & Frackowiak, R. S. (2010). Irritability in pre-clinical Huntington's disease.

*Neuropsychologia*, 48 (2), 549-557. doi: 10.1016/j.neuropsychologia.2009.10.016.

Leibenluft, E., & Stoddard, J. (2013). The developmental psychopathology of irritability.

*Development and Psychopathology*, 25, 1473-1487. doi:

10.1017/S0954579413000722.

\*Litvan, I., Paulsen, J.S., Mega, M.S. & Cummings, J.L. (1998). Neuropsychiatric

assessment of patients with hyperkinetic and hypokinetic movement disorders.

*Archives of Neurology*, 55 (10), 1313-1319. doi: 10.1001/archneur.55.10.1313.

Mangelli, L., Fava, G.A., Grassi, L., Ottolini, F., Paolini, S., Porcelli, P., ... Sonino, N.

(2006). Irritable mood in Italian patients with medical disease. *The Journal of*

*Nervous and Mental Disease*, 194 (3), 226-228. doi:

10.1097/01.nmd.0000202511.21925.a2.

Mays, N., Roberts, E., & Popay, J. (2001). Synthesizing research evidence. In N. Fullop, P.

Allen, A. Clarke, & N. Black (eds.) *Studying the organisation and delivery of health services: Research Methods*. Routledge: London, 188-219.

Mega, M.S., & Cummings, J.L. (1994). Frontal-subcortical circuits and neuropsychiatric

disorders. *Journal of Neuropsychiatry and Clinical Neuroscience*, 6, 358-370. doi:

10.1176/jnp.6.4.358.

\*Nimmagadda, S.R., Agrawal, N., Worrall-Davies, A., Markova, I. & Rickards, H. (2011).

Determinants of irritability in Huntington's disease. *Acta Neuropsychiatrica*, 23, 309-

314. doi: 10.1111/j.1601-5215.2011.00563.x.

## IRRITABILITY IN HUNTINGTON'S DISEASE

- Novak, M.J.U., & Tabrizi, S.J. (2010). Huntington's disease. *BMJ*, *341*, 34-40. doi: 10.1136/bmj.c3109.
- \*Paulsen, J.S., Ready, R.E., Hamilton, J.M., Mega, M.S., & Cummings, J.L. (2001). Neuropsychiatric aspects of Huntington's disease. *Journal of Neurology, Neurosurgery and Psychiatry*, *71*, 310-314. doi: 10.1136/jnnp.71.3.310.
- \*Pflanz, S., Besson, J.A.O., Ebmeier, K.P., & Simpson, S. (1991). The clinical manifestation of mental disorder in Huntington's disease: A retrospective case record study of disease progression. *Acta Psychiatrica Scandinavica*, *83*, 53-60. doi: 10.1111/j.1600-0447.1991.tb05511.x.
- \*Reedeker, N., Bouwens, J.A., Giltay, E.J., Le Mair, S.E., Roos, R.A.C., van der Mast, R.C., & van Duijn, E. (2012). Irritability in Huntington's disease. *Psychiatry Research*, *200*, 813-818. doi: 10.1016/j.psychres.2012.03.041.
- \*Rickards, H., De Souza, J., van Walsem, M., van Duijn, E., Simpson, S.A., Squitieri, F., ... & The European Huntington's Disease Network (2011). Factor analysis of behavioural symptoms in Huntington's disease. *Journal of Neurology, Neurosurgery and Psychiatry*, *82*, 411-412. doi: 10.1136/jnnp.2009.181149.
- Roos, R.A.C. (2014). Clinical Neurology. In G.P. Bates, Tabrizi, S.J., & Jones, L. (eds.) *Huntington's Disease* (4<sup>th</sup> Ed.). Oxford: Oxford University Press.
- Siemer, M. (2009). Mood experience: Implications of a dispositional theory of moods. *Emotion Review*, *1*, 256-263. doi: 10.1177/1754073909103594.
- Snaith, R.P., Constantopoulos, A.A., Jardine, M.Y., & McGuffin, P. (1978). A clinical scale for the self-assessment of irritability. *The British Journal of Psychiatry*, *132*, 164-171. doi: 10.1192/bjp.132.2.164.

## IRRITABILITY IN HUNTINGTON'S DISEASE

- Snaith, R.P., & Taylor, C.M. (1985). Irritability: definition, assessment and associated factors. *British Journal of Psychiatry*, *147*, 127-136. doi: 10.1192/bjp.147.2.127.
- Tabrizi, S. J., Langbehn, D. R., Leavitt, B. R., Roos, R. A., Durr, A., Craufurd, D., ... & Borowsky, B. (2009). Biological and clinical manifestations of Huntington's disease in the longitudinal TRACK-HD study: Cross-sectional analysis of baseline data. *The Lancet. Neurology*, *8* (9), 791-801. doi: 10.1016/S1474-4422(09)70170-X.
- \*Thompson, J.C., Harris, J., Sollom, A.C., Stopford, C.L., Howard, E., Snowden, J.S., & Craufurd, D. (2012). Longitudinal evaluation of neuropsychiatric symptoms in Huntington's disease. *The Journal of Neuropsychiatry and Clinical Neurosciences*, *24* (1), 53-60. doi: 10.1176/appi.neuropsych.11030057.
- \*Thompson, J.C., Snowden, J.S., Craufurd, D., & Neary, D. (2002). Behavior in Huntington's disease: Dissociating cognition-based and mood-based changes. *The Journal of Neuropsychiatry and Clinical Neurosciences*, *14* (1), 37-43. doi: 10.1176/jnp.14.1.37.
- \*Van den Stock, J., De Winter, F-L., Ahmad, R., Sunaert, S., Van Laere, K., Vandenberghe, W., & Vandenberghe, M. (2015). Functional brain changes underlying irritability in premanifest Huntington's disease. *Human Brain Mapping*, *36*, 2681-2690. doi: 10.1002/hbm.22799.
- \*Van Duijn, E. (2010). Treatment of irritability in Huntington's disease. *Current Treatment Options in Neurology*, *12*, 424-433. doi: 10.1007/s11940-010-0088-3.
- \* Van Duijn, E., Craufurd, D., Hubers, A.A.M., Giltay, E.J., Bonelli, R., Rickards, H., ... & the European Huntington's Disease Network Behavioural Phenotype Working Group. (2014). Neuropsychiatric symptoms in a European Huntington's disease cohort

## IRRITABILITY IN HUNTINGTON'S DISEASE

(REGISTRY). *Journal of Neurology, Neurosurgery and Psychiatry*, 85, 1411-1418.

doi: 10.1136/jnnp-2013-307343.

Van Duijn, E., Kingma, E.M., & van der Mast, R.C. (2007). Psychopathology in verified Huntington's disease gene carriers. *Journal of Neuropsychiatry and Clinical Neuroscience*, 19 (4), 441-448. doi: 10.1176/appi.neuropsych.19.4.441.

\*Van Duijn, E., Reedecker, N., Giltay, E.J., Eindhoven, D., Roos, R.A.C., & van der Mast, R.C. (2013). Course of irritability, depression and apathy in Huntington's disease in relation to motor symptoms during a two-year follow up period. *Neurodegenerative Diseases*. doi: 10.1159/000343210.

\*Vassos, E., Panas, M., Kladi, A., & Vassilopoulos, D. (2007). Higher levels of extroverted hostility detected in gene carriers at risk for Huntington's disease. *Biological Psychiatry*, 62, 1347-1352. doi: 10.1016/j.biopsych.2006.12.016.

Wagle, A.C., Wagle, S.A., Markova, L.S., & Berrios, G.E. (2000). Psychiatric morbidity in Huntington's disease: The current view. *Neurology, Psychiatry and Brain Research*, 8, 5-16.

Note: \* indicates inclusion in the review.

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**Table 1***Summary of studies of irritability in HD*

<b>Citation</b>	<b>Participants (N)</b>	<b>Gender (N)</b>	<b>Age (Mean)</b>	<b>HD stage</b>	<b>Irritability measures</b>	<b>Other measures</b>
Banaszkiewicz et al. (2012)	HD patient-caregiver dyads (80)	-	47.7	-	UHDRS-b	HAM-D
Berrios et al. (2001)	HD (26)	Female (10) Male (16)	37.8	People with HD	IRR	PER, BDI, CFQ, SIGNAL, MOC, DIS, STAI & STAI2
Berrios et al. (2002)	Gene carriers (32) Non carriers (66)	Female (56) Male (42)	46.7	Asymptomatic	IRR	PER, BDI, CFQ, SIGNAL, MOC, DIS
Bouwens et al. (2015)	Mutation carriers (90)	Female (49) Male (41)	49	Pre-motor symptomatic (25) Motor symptomatic (64)	Irritability Scale (Chatterjee)	PBA UHDRS
Burns et al. (1990)	HD Gene carriers (26) Alzheimer's disease (31)	Female (29) Male (28)	48.3 (HD) 70.3 (AD)	-	Irritability/Apathy Scale (developed for this research)	Yudofsky Aggression Scale
Chatterjee et al. (2005)	Gene carriers (53) Caregivers (53)	Female (21) Male (32)	48.2	-	John Hopkins Irritability Questionnaire	BDI Apathy Scale MMSE
Craufurd et al. (2001)	Gene carriers (134)	Female (71) Male (63)	50	Various	UHDRS, PBA-HD	
Gregory et al (2015)	Gene carriers (45) Pre-manifest HD (39)	Female (49) Male (35)	46	Pre-symptomatic (39) Early symptomatic (45)	PBA	HADS
Groves et al. (2011)	Physician leaders from HD (55) speciality centres	-	-	-	-	-

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	(55)					
Hubers et al. (2013)	Gene carriers (2106 at baseline, 945 at follow-up)	Female (1034) Male (1072)	50.3	Motor symptomatic	UHDRS-b	-
Julien et al. (2007)	Gene carriers (89) Non carriers (115)	Female (123) Male (81)	38	-	CIDI	-
Kingma et al. (2008)	Non-carriers (56) Gene carriers (152)	Female (114) Male (94)	45.3	Pre-symptomatic gene carriers (55) Early symptomatic (47) Advanced symptomatic (50)	PBA	UHDRS-m
Kirkwood et al. (2002a)	Gene carriers (12) Non-carriers (31)	Female (28) Male (15)	44	Pre-symptomatic (12) Non-carriers (31)	Abbreviated MMPI (irritability scale)	-
Kirkwood et al. (2002b)	HD (175) Non carriers (363)	Female (384) Male (154)	41.4	Pre-symptomatic (149) Manifest HD (26)	Abbreviated MMPI Irritability scale (content analysis of MMPI items)	-
Kloppel et al. (2010)	Gene carriers (16) Controls (15)	Female (16) Male (15)	39.3 40.4	Pre-symptomatic	SIS, John Hopkins Irritability Questionnaire	BDI BIS-11 STAI
Litvan et al. (1998)	HD (29) Progressive Supranuclear Palsy (34)	-	43.8 (HD) 66.6 (PSP)	Various stages	NPI	UHDRS
Nimmagadda et al. (2011)	PwHD & their carers (30)	Female (14) Male (16)	49.17	Genetically confirmed HD	IDA BIS	BADS MADRS UHDRS-m STAI

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Paulsen et al. (2001)	HD (52) Caregivers (52)	Female (27) Male (25)	45.5	Various	NPI	UHDRS
Pflanz et al. (1991)	HD (86)	HD: Male (17) Female (20) Deceased: Male (17) Female (32)		Various	Present State Examination (9 <sup>th</sup> Ed.)	-
Reedeker et al. (2012)	Gene carriers (130) Non carriers (43) Informants (158)	-	-	-	IS PBA UHDRS-b	UHDRS-m CIDI
Rickards et al. (2010)	People with HD (1690)	-	-	-	UHDRS-b	-
Thompson et al. (2002)	People with HD (82)	Female (41) Male (41)	49	Clinically diagnosed HD	PBA-HD UHDRS-b	-
Thompson et al. (2012)	HD (111)	Female (68) Male (43)	48	Clinically diagnosed HD	PBA-HD	-
Van den Stock et al. (2015)	Gene carriers (20) Non carriers (20)	Female (23) Male (17)	37.5	Pre-manifest	PBA-HD	UHDRS BDI STAI
Van Duijn. (2010)	Review of treatment studies	-	-	-	-	-
Van Duijn et al. (2013)	HD (121)	-	-	Pre-symptomatic = 46 Symptomatic = 75	PBA	-
Van Duijn et al. (2014)	Gene carriers (1993)	Female (977) Male (1016)	50.3	Early and mid-stage	UHDRS-b	-
Vassos, Panas, Kladi & Vassilopoulos (2007)	Gene carriers (29) Non-carriers (35)	Female (37) Male (27)	34.2	-	UHDRS SIS HDHQ	MOC

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Note: AD = Alzheimer's disease; BADS = Behavioural Assessment of Dysexecutive Syndrome; BDI = Beck Depression Inventory; DIS = Dissociation Questionnaire; CFQ = Cognitive Failures Questionnaire; CIDI = Composite International Diagnostic Interview; HAM-D = Hamilton Depression Rating Scale; HDHQ = Hostility & Direction of Hostility Questionnaire; IRR = Snaith's Irritability Scale; IS = Irritability Scale; MADRS = Montgomery & Asberg Depression Rating Scale; MMPI = Minnesota Multiphasic Personality Inventory; MMSE = Mini-Mental State Exam; MOC = Maudsley Obsessive-Compulsive Questionnaire; NPI = Neuropsychiatric Inventory; PBA = Problem Behaviours Assessment; PER = Personality Deviance Scale; SIGNAL = Signal Detection Memory Test; SIS = Snaith Irritability Self-Assessment Scale; STAI & STAI2 = Spielberger Anxiety scales; UHDRS = Unified Huntington's Disease Rating Scale

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**Table 2***Results of studies of irritability in HD*

<b>Citation</b>	<b>Aim</b>	<b>Results</b>
Banaszkiewicz et al. (2012)	To identify determinants of quality of life, functional disability and caregiver burden.	Irritability is not significantly associated with disability.
Berrios et al. (2001)	To investigate the relationship between psychiatric profile and CAG repeats.	Compared with available norms, participants showed increased levels of 'outward irritability'. No significant correlation with irritability and CAG repeat length.
Berrios et al. (2002)	To compare psychiatric profiles of gene carriers and non-carriers.	Significant difference in inward and outward irritability between GC and NC, with irritability being higher in GC. Factor structure: inward and outward irritability were included within the 'personality' factor.
Bouwens et al. (2015)	To investigate the course and temporal relationship between irritability and other psychological difficulties.	No significant increase in irritability from baseline to follow-up. At baseline 33% of people with HD were irritable, with 70% of those remaining irritable at 2-year follow-up. Of those who were not irritable at baseline 23% developed irritability at 2-year follow-up. Multivariate regression model showed an association between increase in apathy and an increase in irritability. Continuous use of antipsychotics associated with an increase in irritability.
Burns et al. (1990)	To compare irritability, aggression and apathy in people with HD with people with AD.	No significant difference in irritability or apathy between the HD and AD groups. HD group were significantly more aggressive than the AD group and aggressive outbursts lasted longer in the HD group. Irritability, apathy & aggression were independent of each other in both groups. Irritability correlated positively with bad temper in the HD group but there was no correlation in the AD group.
Chatterjee et al. (2005)	To examine agreement between people with HD and their caregivers regarding presence of irritability, apathy and depression.	No significant difference in report of irritability between PwHD and caregivers. No difference in BDI scores. Difference in apathy scores between the two groups.

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Craufurd et al. (2001)	To understand behavioural abnormalities in HD and develop a method of assessing these changes.	Irritability present in 44% of sample (severity rating of 2 or more). Three factors obtained from factor analysis: 1 - apathy; 2 - irritability; 3 - depression. Irritability difficulties occurred more frequently in people with HD with an illness duration of 6-11 years. Irritability factor showed no correlation with duration of illness or CAG repeat length.
Gregory et al (2015)	Investigate structural connectivity and changes associated with depression, apathy and irritability in HD.	Significant difference in irritability between the two groups. Significant negative correlations between irritability score and fractional anisotropy which was dependent on cumulative probability to onset.
Groves et al. (2011)	To provide direction for the management of irritability in HD.	SSRIs were most frequently used to treat mild to moderate irritability in HD. Antipsychotics (APD) were more commonly used in Europe to treat mild to moderate irritability than in North America & Australia. SSRIs used when irritability occurred with comorbid depression and anxiety. APDs used when irritability occurred with aggression and impulsivity.
Hubers et al. (2013)	To investigate predictors and correlates of suicidal ideation in HD.	Baseline presence of irritability significantly correlated with suicidal ideation – those with suicidal ideation were more irritable than those without. Multivariate analyses indicated irritability was not an independent correlate of suicidal ideation. At follow-up, irritability was not a predictor of suicidal ideation in HD.
Julien et al. (2007)	To compare the prevalence of psychological difficulties in pre-symptomatic gene carriers and non-carriers and to look at the relationship with proximity to onset.	Gene carriers reported a greater prevalence of 'manic' symptoms (11%) compared with NGC (4%) – in every case irritability was reported. Irritability was increased in gene carriers up to 10 years prior to clinical onset but not in those further from onset. No significant relationship between proximity to onset and irritability within the 10 year period.
Kingma et al. (2008)	To investigate the behavioural difficulties in HD.	Factor analysis revealed 3 components: irritability, apathy and depression. All mutation carriers showed significantly more irritability, apathy & depression than non-carriers. No significant difference in irritability between ASGC and other disease stages. No significant relationship between irritability and depression or apathy.
Kirkwood et al. (2002a)	To examine whether longitudinal changes in personality can be detected in pre-symptomatic gene carriers.	Greater increase irritability and clinical hostility observed over time in the PSGC group compared with NGC. No correlation between number of CAG repeats and irritability in both groups.

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Kirkwood et al. (2002b)	To investigate whether psychological difficulties can be detected in pre-symptomatic HD.	No significant difference in MMPI scores across groups. No significant difference in irritability across the three groups and no association with proximity to onset.
Kloppel et al. (2010)	To examine the emotional neurocircuitry associated with irritation,	No significant difference in irritability between PSC and controls. Companions ratings did not differ from those of the PSC. Ratings on the SIS were within normal range apart from 1 PSC. Negative emotions positively correlated with SIS & BIS-11.
Litvan et al. (1998)	To compare neuropsychiatric aspects of HD compared with PSP.	Irritability influenced the total NPI score in PwHD. PwHD scored significantly higher on agitation, irritability and anxiety while those with PSP scored higher for apathy. In PwHD, agitation was correlated with anxiety, irritability, disinhibition and euphoria. Irritability was associated with anxiety, disinhibition, euphoria and depression. Logistic regression analysis indicated PwHD are more likely to exhibit hyperactive behaviour. People with PSP are more likely to exhibit hypoactive behaviour.
Nimmagadda et al. (2011)	To investigate the association of irritability in HD with other psychological constructs and movement disorder.	Both inward and outward irritability were significantly positively associated with MADRS scores, STAI state and trait anxiety scores. BIS scores were positively associated with STAI trait scores and both outward and inward irritability scores on the IDA. Negative correlation between irritability scores and the UHDRS.
Paulsen et al. (2001)	To use the NPI to characterise neuropsychiatric symptoms in HD.	Irritability endorsed in 65.4% of sample. NPI - High correlation between irritability & agitation indicating two scales are measuring the same construct. Irritability also correlated with anxiety and disinhibition.
Pflanz et al. (1991)	To determine the range and frequency of psychological difficulties in HD.	Irritability present in 64% of cases and was the 2 <sup>nd</sup> most common difficulty. Irritability occurred between 0-3 years prior to onset of motor symptoms. Loss of interest and concentration correlated with irritability.
Reedeker et al. (2012)	To investigate the psychometric properties of the Irritability Scale against the PBA irritability factor to establish a reliable cut off.	Irritability significantly higher in MC (35% irritable) than NC (9% irritable). 28% of MC considered irritable according to IS-self and informant scales. 50% considered not irritable according to both scales. For the remaining 23% there was disagreement between participants and informants (18/27 reported selves as not irritable but their informant did). Irritability independently correlated with benzodiazepine use.
Rickards et al. (2010)	To perform a factor analysis on completed UHDRS-b	Factor analysis indicated that irritability is a distinct 'psychiatric symptom' in HD.

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	assessments.	
Thompson et al. (2002)	To investigate how behavioural change in HD relates to other indices of disease severity.	Depression & irritability subscales poorly correlated with functional capacity, motor impairment & cognition. Apathy was significantly correlated. UHDRS-b score significantly correlated with PBA-HD depression & irritability subscales. UHDRS irritability scale significantly correlated with irritability subscale of the PBA-HD.
Thompson et al. (2012)	To evaluate the prevalence of neuropsychiatric difficulties in HD over time.	Irritability common with a prevalence ranging from 49-83%. Longitudinal analysis showed an increase in irritability over time with a significant linear effect in those who entered the study at stage I and II but not in those who entered at stage III of HD.
Van den Stock et al. (2015)	Identify structural and functional brain changes underlying irritability in pre-manifest HD.	Irritability significantly higher in GC vs NC.
Van Duijn. (2010)	To review the treatments of irritability.	Suggests use of an SSRI as a first choice medication to manage irritability in HD or a mood stabiliser. An alternative would be an antipsychotic. Behavioural or other psychotherapeutic interventions should be considered.
Van Duijn et al. (2013)	To investigate the progression of irritability, depression and apathy in HD over a 2-year follow up.	2-year follow-up: No significant change in irritability. Associations between PBA factor scores and UHDRS-m: as UHDRS-m score increased so did the PBA irritability factor. In pre-symptomatic group, strongest relationship was between an increased UHDRS-m score and increased irritability score. At follow-up 15 of the pre-symptomatic group were symptomatic. No significant increase in irritability compared with those who remained pre-symptomatic.
Van Duijn et al. (2014)	To examine the occurrence and correlates of neuropsychiatric symptoms in HD.	61.4% of HD mutation carriers scored 'no irritability', 24.7% scored 'mild irritability' and 13.9% scored 'moderate/severe irritability'. The prevalence of moderate/ severe irritability increased by stage of disease from 10.4% at stage 1 to 19.6% at stages 4-5. Irritability independently correlated with male sex, younger age, a history of depression, psychosis and a previous suicide attempt.
Vassos, Panas, Kladi & Vassilopoulos (2007)	To distinguish which behavioural and psychiatric features differentiate gene carriers with non-carriers.	No significant difference in irritability between GC and NC. Higher extroverted hostility in GC than in NC. Overlap between the two groups suggests extroverted hostility may not be pathologic in GC.

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**Table 3***Measures of irritability in HD*

<b>Measure</b>	<b>Description</b>	<b>Reliability</b>	<b>Validity</b>
Burns Irritability Scale (BIS; Burns, Folstein, Brandt & Folstein, 1990)	Measures irritability and apathy according to carer's ratings and does not include subjective experience. It uses a 5-point scale assessing the presence of irritability ranging from "never" to "always".	Internal consistency: - Irritability: $\alpha = 0.82$ - Apathy: $\alpha = 0.78$ Inter-rater <sup>1</sup> : - Whole interview: $\kappa = 0.98$ - Irritability: $\kappa = 1.00$ - Apathy: $\kappa = 0.85$ Test-retest: - Whole interview: $\kappa = 0.88$ - Irritability: $\kappa = 0.81$ - Apathy: $\kappa = 0.76$	Convergent: - Psychogeriatric Dependency Rating Scale: $r = 0.87$
Irritability, depression, anxiety scale (IDA; Snaith, Constantopoulos, Jardine & McGuffin 1978)	Scale assessing irritability, depression and anxiety to be used within clinical context. Irritability understood as a temporary psychological state. Includes 8 irritability items	Inter-rater: - Outward irritability: $r = .87-.90$ - Inward irritability: $r = .74-.90$ - Depression: $r = .80-.90$ - Anxiety: $r = .75-.80$ Split-half: - Outward irritability: $r = .77, .80, .88$ - Inward irritability: $r = .70, .92, .93$ - Depression: $r = .72, .77, .81$ - Anxiety: $r = .74, .80, .87$	
Irritability Questionnaire (IRQ; Craig, Hietenan,	Subjective measure of irritability. Consists of 21	Internal consistency: - Global: $\alpha = 0.90$	Convergent: - Trait anger scale: $r = 0.72$

<sup>1</sup> Looked at the presence or absence of irritability.

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Markova & Berrios, 2008)	items assessing the frequency and severity of irritability with each individual item score ranging from 0-3.	Split half = 0.78 - Frequency: $\alpha = 0.90$ Split half = 0.77 - Severity: $\alpha = 0.89$ Split half = 0.58 Retest reliability: $r = 0.82$	- State anger scale: $r = 0.58$ - IDA outward: $r = 0.58$ - IDA inward: $r = 0.49$ - BIS: $r = 0.37$
John Hopkins Irritability Scale (Chattergee, Anderson, Moskoqitz, Hauser & Marder, 2005)	Objective measure (informant-report) of irritability. Consists of 14 items pertaining to irritability with the range of all possible scores being 0-42 to assess the presence of irritability.	No data available	No data available
Problem Behaviours Assessment – Huntington's disease (PBA-HD; Craufurd, Thompson & Snowden, 2001)	Semi-structured interview measuring behavioural difficulties in HD including the presence, severity and frequency.	Inter-rater: - Severity: $r = 0.86$ - Frequency: $r = 0.84$ Internal consistency: $\alpha = 0.67$ Test-retest: - Severity: $r = 0.94$ - Frequency: $r = 0.92$	
Unified Huntington's Disease Rating Scale (UHDRS; Huntington Study Group, 1996)	Assesses difficulties in motor, cognitive, functional and behavioural domains. The behavioural section measures the frequency and severity of difficulties related to affect, thought content and coping styles.	Internal Consistency: - Behavioural: $\alpha = 0.83$ - Motor: $\alpha = 0.95$ - Cognitive: $\alpha = 0.90$ - Functional: $\alpha = 0.95$	Divergent (Behavioural Total): - Motor: $r = -0.10$ - Total Functional Capacity: $r = -0.07$

## Appendix 1-A: Author Guidelines

### British Journal of Health Psychology

#### Author Guidelines

The aim of the British Journal of Health Psychology is to provide a forum for high quality research relating to health and illness. The scope of the journal includes all areas of health psychology as outlined in the Journal [Overview](#).

The types of paper invited are:

- papers reporting original empirical investigations, using either quantitative or qualitative methods, including reports of interventions in clinical and non-clinical populations;
- theoretical papers which may be analyses or commentaries on established theories in health psychology, or presentations of theoretical innovations;
- we particularly welcome review papers, which should aim to provide systematic overviews, evaluations and interpretations of research in a given field of health psychology; and
- methodological papers dealing with methodological issues of particular relevance to health psychology.

All papers published in The British Journal of Health Psychology are eligible for Panel A: Psychology, Psychiatry and Neuroscience in the Research Excellence Framework (REF).

#### 1. Circulation

The circulation of the Journal is worldwide. Papers are invited and encouraged from authors throughout the world.

#### 2. Length

Papers should normally be no more than 5000 words (excluding the abstract, reference list, tables and figures), although the Editor retains discretion to publish papers beyond this length in cases where the clear and concise expression of the scientific content requires greater length.

#### 3. Editorial policy

The Journal receives a large volume of papers to review each year, and in order to make the process as efficient as possible for authors and editors alike, all papers are initially examined by the Editors to ascertain whether the article is suitable for full peer review. In order to qualify for full review, papers must meet the following criteria:

- the content of the paper falls within the scope of the Journal
- the methods and/or sample size are appropriate for the questions being addressed
- research with student populations is appropriately justified
- the word count is within the stated limit for the Journal (i.e. 5000 words)

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### 4. Submission and reviewing

All manuscripts must be submitted via [Editorial Manager](#). The Journal operates a policy of anonymous (double blind) peer review. We also operate a triage process in which submissions that are out of scope or otherwise inappropriate will be rejected by the editors without external peer review to avoid unnecessary delays. Before submitting, please read the [terms and conditions of submission](#) and the [declaration of competing interests](#). You may also like to use the [Submission Checklist](#) to help you prepare your paper.

### 5. Manuscript requirements

- Contributions must be typed in double spacing with wide margins. All sheets must be numbered.
- Manuscripts should be preceded by a title page which includes a full list of authors and their affiliations, as well as the corresponding author's contact details. A template can be downloaded from [here](#).
- For articles containing original scientific research, a structured abstract of up to 250 words should be included with the headings: Objectives, Design, Methods, Results, Conclusions. Review articles should use these headings: Purpose, Methods, Results, Conclusions. As the abstract is often the most widely visible part of your paper, it is important that it conveys succinctly all the most important features of your study. You can save words by writing short, direct sentences. Helpful hints about writing the conclusions to abstracts can be found [here](#).
- Statement of Contribution: All authors are required to provide a clear summary of 'what is already known on this subject?' and 'what does this study add?'. Authors should identify existing research knowledge relating to the specific research question and give a summary of the new knowledge added by your study. Under each of these headings, please provide 2-3 (maximum) clear outcome statements (not process statements of what the paper does); the statements for 'what does this study add?' should be presented as bullet points of no more than 100 characters each. The Statement of Contribution should be a separate file.
- Conflict of interest statement: We are now including a brief conflict of interest statement at the end of each accepted manuscript. You will be asked to provide information to generate this statement during the submission process.
- The main document must be anonymous. Please do not mention the authors' names or affiliations (including in the Method section) and always refer to any previous work in the third person.
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Author, A., Author, B., & Author, C. (1995). *Title of book*. City, Country: Publisher.

Author, A. (2013). Title of journal article. *Name of journal*, 1, 1-16. doi: 10.1111/bjep.12031

- SI units must be used for all measurements, rounded off to practical values if appropriate, with the imperial equivalent in parentheses.
- In normal circumstances, effect size should be incorporated.
- Authors are requested to avoid the use of sexist language.
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**Section 2: Research Paper**

**Understandings of psychological difficulties in people with Huntington's disease and their expectations of psychological therapy**

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**Abstract**

**Aim:** This study sought to investigate how people with Huntington's disease (HD) understand and experience psychological distress in HD and their expectations of psychological therapy.

**Method:** A qualitative methodology was adopted involving semi-structured interviews and interpretative phenomenological analysis (IPA). A total of nine participants (five women and four men) who had opted in to engage in a trial of mindfulness-based cognitive therapy (MBCT) were recruited and interviewed prior to the MBCT trial. Interviews were transcribed verbatim and analysed using IPA whereby themes were analysed within and across transcripts and classified into superordinate themes.

**Results:** Three superordinate themes were developed: Attributing psychological distress to HD: *"you're blaming everything on that now"*; Changes across time: *"in the past you'd just get on with it"*; Therapy instils hope and fight: *"a light at the end of the tunnel"*.

**Conclusion:** Understandings of psychological distress in HD ranged from biological to psychological explanations, with both often being accepted simultaneously by the same individual. Individual experience seemed to reflect a dynamic process whereby people's understanding and experience changed over time. Psychological therapy was accepted as a positive alternative to medication, providing people with HD with hope that their psychological wellbeing could be enhanced.

**Keywords:** Huntington's disease; psychological difficulties; psychological therapy, mindfulness based cognitive therapy

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Understandings of psychological difficulties in people with Huntington's disease and their expectations of psychological therapy

Huntington's disease (HD) is a chronic neurodegenerative disease which causes problems with movement, coordination, cognitive functioning, and is often also associated with a number of different emotional difficulties. It is suggested that around five to ten per 100,000 people are affected (Kay, Fisher & Hayden, 2014) and as HD is a genetic disease with a 50% chance of inheriting the affected gene from a parent with HD, people with HD have often seen their parents affected by the disease (Kremer, 2002). People are generally diagnosed between the ages of 35-55 years with a life expectancy of around 15-20 years after diagnosis (which is usually given upon the onset of motor symptoms; Keenan, Simpson, Miedzybrodzka, Alexander & Semper, 2013). Considering the age at which people may be diagnosed with HD it may be reasonable to view this as a 'disruptive event' (Bury, 1982) in which people are required to re-evaluate the trajectory of their life and attempt to adjust accordingly.

For people with a family history of HD, and who are subsequently at risk, predictive testing can be carried out prior to an individual showing any symptoms (Novak & Tabrizi, 2010). This will indicate whether or not a person will go on to develop HD in the future. Additionally, a diagnostic test is performed once a person presents with problems indicative of HD (Novak & Tabrizi, 2010). It is important to consider the psychological and emotional implications attached to accessing these tests and the results, given that someone receiving a positive test will go on to develop HD (Meiser & Dunn, 2000). For an individual who receives a positive predictive test, they are left knowing there is no cure but with the uncertainty of when and how the disease will begin to progress. However, some people find this uncertainty more tolerable than the uncertainty of not knowing whether they have inherited the affected gene (Novak & Tabrizi, 2010).

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People with HD often experience emotional difficulties. The most common include depression, anxiety, apathy and irritability (Kirkwood, Su, Conneally & Foroud, 2001) and these have the potential to impact on quality of life, perhaps even more so than motor problems or cognitive impairment (Ho, Gilbert, Mason, Goodman & Barker, 2009). It has been argued that difficulties with mood, such as depression, are often one of the earliest signs of HD preceding motor difficulties (Pla, Orvoen, Saudou, David & Humbert, 2014). High levels of depression in HD, ranging from 33-69% (see van Duijn, Kingma & van Der Mast, 2007 for review), have become expected as a result of the understanding it has a biological origin, the potential for cognitive impairment and the 50% risk of passing the gene on to children (Paulsen et al., 2005). Moreover, depression is observed most often in HD when the HD starts to impact on an individual's functional capacity and independence (Paulsen et al., 2005).

In addition, anxiety also co-occurs alongside depression. In a systematic review, Dale and van Duijn (2015) found that anxiety was present in between 13% to 71% of people with manifest HD. Additionally, there was no significant difference between people with manifest (presence of motor symptoms) and pre-manifest (confirmation of HD gene but motor symptoms currently absent) HD in levels of anxiety. The presence of anxiety in HD may be a result of environmental stressors whereby people may become overwhelmed by their situation as well as tasks that may have previously required little attention (Hoffman, 1999). Indeed, elevated levels of anxiety were found to be present in those who were gene positive, both close to and far from onset (Duff, Paulsen, Beglinger, Langbehn & Stout, 2007).

Furthermore, irritability is commonly reported in people with HD and has been shown to be present in up to 50 percent of people with HD (Craufurd, Thompson & Snowden, 2001; Dewhurst, Oliver, Trick & McNight, 1969). Indeed, irritability, alongside anxiety and depression, is noted to be a core psychological feature of HD at the pre-symptomatic stage

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(Kloppel et al., 2010); it is argued to cause significant distress, not only to the person with HD but to those around them such as family members and carers (Nimmagadda, Agrawal, Worrall-Davies, Markova & Rickards, 2011). However, Kingma et al. (2008) found that depression and irritability are not linked to stage of disease with similar levels found in those at pre-, early and advanced stages indicating that people with HD may experience these for many years and subsequently need support throughout the progression of the disease. Similarly, Nimmagadda et al., (2011) suggested that irritability was related to the behavioural and affective difficulties in HD rather than the progressive motor and cognitive difficulties.

The dominant perspectives within the HD field are to look for largely biological determinants of distress. For example, many researchers take the view that psychological difficulties occur as a result of biological factors whereby neural mechanisms in the brain are affected by HD which then subsequently affect mood (Paulsen et al., 2005). Indeed, Kowalski, Belcher, Keltner and Dowben (2015) summarised that depression, one of the most common psychological difficulties in HD, “appears to be a direct neurological consequence of the brain condition, rather than a psychological reaction to this serious illness” (p.159).

However, psychological distress can also be understood from different perspectives within the broader field of chronic illness research and this is starting to influence how distress in HD can be understood. For example, evidence suggests that psychological factors such as what people believe about the illness and coping strategies are also influential in predicting psychological distress and well-being in people with HD (Arran, Craufurd & Simpson, 2013; Kaptein et al., 2006). A reaction to the onset of a disease such as HD may explain the occurrence of depression in HD (Pla et al., 2014). Similarly, Julien et al. (2007) also proposed that difficulties such as depression are reactive and indicate an emotional response to the awareness of future motor impairment.

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Perhaps not surprisingly given the dominance of biological accounts, medication such as anti-depressants are often used to manage psychological difficulties in HD (Craufurd & Snowden, 2011). However, regardless of its efficacy, medication may not always be the preferred option for people with HD as they have to manage the potential side effects of medication (Aubeeluck & Wilson, 2008) and how this can impact on their own drug regimes. Consequently, psychological interventions may provide an alternative or additional way to reduce distress.

Currently, little evidence exists of the acceptability and efficacy of psychological approaches for people with HD. However, in another neurodegenerative disease, Parkinson's disease (PD), there is increasing evidence to support the use of psychological interventions for low mood and anxiety in this population (see Charidimou, Seamons, Selai & Schrag, 2011, for a review). Such approaches include mindfulness-based cognitive therapy (MBCT; Fitzpatrick, Simpson & Smith, 2010) and cognitive behavioural therapy (CBT; Dobkin et al., 2011). However, currently no trial has been conducted evaluating a psychological therapy in people with HD. Clearly, then it could be useful to assess whether psychological interventions are seen as potentially beneficial.

As a result, this study adopted a qualitative methodology in order to obtain detailed accounts of people with HD's understanding and experience of psychological difficulties and expectations of psychological therapy. Given the dominance of biological accounts for psychological problems, at least within the scientific and clinical community, it was considered important to understand whether beliefs about cause of distress and the possibility of therapy would be consistent. In order to address this, semi-structured interviews were conducted and analysed using interpretative phenomenological analysis (IPA; Smith & Osborn, 2003). Due to the inductive nature of this analytic approach it was possible to gain

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insight into the lived experiences of individuals and as such is appropriate to understand how people with HD perceive and experience psychological distress and psychological therapy.

Consequently, this study aimed to investigate individuals with HD's understanding of psychological difficulties in HD and their views of psychological therapy. Participants were recruited from those due to take part in a trial of MBCT and therefore the study focused on people's knowledge of psychological therapy, as well as their hopes and expectations of a psychological approach.

### **Method**

#### **Design**

The study employed a qualitative methodology to obtain participants' understanding of psychological distress and the opportunities offered by psychological therapy in the context of HD. IPA (Smith & Osborn 2003) was used to analyse the data. IPA is widely used in psychological research and aims to explore how people understand and make sense of their experiences within their personal, and social world (Smith, Flowers & Larkin, 2009).

#### **Participants**

People with HD were eligible to take part. Participants with HD were recruited from an ongoing MBCT trial (Clinicaltrials.gov: NCT02464293) of which a summary can be seen in Appendix 2-A. To be included in the MBCT trial participants had to meet the following criteria: confirmed CAG expansion on the huntingtin gene; be a gene carrier and either be pre-symptomatic or at an early stage; have clinical signs of low mood or depression as identified in their clinical notes or other information recorded at their last clinic visit; be aged 18 years or over; and have not had any changes in their medication six weeks prior to the start of the MBCT trial. Participants were excluded if they had current active suicidal intent.

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In the current study all participants were required to understand and be able to speak English and be aged 18 or over. Nine participants, out of a potential 11, agreed to take part. All participants were people with the HD gene and pre-symptomatic (prior to the onset of motor symptoms), five of whom were female and four male. Participants were aged between 24-56 years with the time since receiving confirmation of the HD gene ranging from 1-17 years. Additionally, six participants were taking antidepressant medication and four participants had previous experience of psychological therapy.

### **Recruitment**

Recruitment took place prior to participants commencing the MBCT course. MBCT is an eight-week group therapy developed by Segal, Williams and Teasdale (2002) which teaches mindfulness skills through a range of practices with the aim of preventing the reoccurrence of depression (Gu, Strauss, Bond & Cavanagh, 2015). Currently, MBCT is being piloted for individuals with HD with the aim of alleviating psychological distress.

Potential participants were initially introduced to the research face to face via a member of the research team from the MBCT trial. Participants were provided with a participant information pack which included a participant information sheet (see Ethics Appendix 4-4) and consent to contact form (see Ethics Appendix 4-5) with a cover letter (see Ethics Appendix 4-6). Participants interested in taking part completed the consent to contact form during their initial meeting with the research team member for the MBCT trial. They were then contacted, either by phone or email depending on their preferred method of contact, by the principal researcher to discuss the study and consider if they would like to take part.

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### **Data Collection**

Data were collected via interviews guided by a semi-structured interview schedule (see Ethics Appendix 4-2). However, further questions were asked which were sensitive to, and guided by, the participants' responses. All interviews were completed during October 2015. All interviews were conducted face to face and at a non-NHS location at a time convenient for the participant. Interviews lasted between 45 and 65 minutes, with an average duration of 54 minutes. At the beginning of each interview the principal researcher checked that each participant had read the participant information sheet and went through the consent form (see Ethics Appendix 4-7), offering participants the chance to ask any questions prior to consenting to participate. All participants signed the consent form to participate and have their interview audio recorded.

Prior to commencing the interview, the principal researcher explained the concept of confidentiality, and its limits, to each participant and ensured they understood this. At the end of each interview participants were debriefed and given time to ask any questions they had about the interview process and subsequent analysis and write up. Each participant was given a pseudonym to retain their anonymity.

### **Data analysis**

All interviews were transcribed verbatim by the principal researcher and all personal identifying information was removed. The interview transcripts were analysed using IPA, following the stages outlined by Smith and Osborn (2003). IPA enables themes to be drawn from the data to reflect the phenomenological understanding participants have of their experiences and the meaning they ascribe to these (Smith & Osborn, 2003). For each participant, their transcript was read then re-read with interesting comments relevant to the research question being noted and used to develop emerging themes. An extract of a

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participant's transcript with annotations and developing themes can be seen in Appendix 2-B.

Following this, emerging themes were then clustered together based on their apparent similarities by copying the emerging themes into a table and giving each cluster a theme name. This was done individually for each participant. Once this was complete, super-ordinate themes were developed which best fit participants' experiences.

### **Ethics**

The study was reviewed and approved by the National Research Ethics Service (NRES) and research governance approval obtained from the relevant hospital trust research and development department.

### **Reflexivity**

In order to ensure the quality of the of qualitative research, principles such as sensitivity to context, rigour and transparency are important to consider (Yardley, 2000) and attempts were made, in both the methodology and reporting of results, to adhere to these principles. The principal researcher attended the MBCT group which enabled a more in depth understanding of participants lives and as such the context in which they made sense of their experiences. Additionally, as a result of attending the MBCT group a reflective diary was kept throughout the research to support the process of reflexivity and ensure the interpretations made were representative of the clients' experiences. This noted information obtained within the group, not provided by participants during the interviews, was bracketed as much as possible to ensure transparency in what information was drawn on when interpreting the data.

Furthermore, the researcher's theoretical position is also important to consider. My epistemological stance is that of a critical realist which assumes that the data gathered provides us with an understanding of a phenomenon but that this is not a direct mirroring,

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rather it is influenced by both the context of the participant and context of the researcher (Harper, 2011).

### Results

Analysis of the data resulted in the development of three themes: Attributing psychological distress to HD: *“you’re blaming everything on that now”*; Changes across time: *“in the past you’d just get on with it”*; Therapy instils hope and fight: *“a light at the end of the tunnel”*.

#### **Attributing psychological distress to HD: *“you’re blaming everything on that now”***

All participants appeared to attribute their psychological distress to HD; however this was from both a biological and psychological perspective insofar as they acknowledged the potential contribution of both biological and psychological factors in causing their distress. Most participants described, more fully, a biological understanding of psychological difficulties in HD. It seemed as though people understood difficulties such as low mood, anxiety and irritability as being part of the disease, resulting from brain changes that occurred due to HD: “I just assumed it’s because with Huntington’s it’s something that’s you know thought will happen...so it was just a case of treating the depression as a biological thing” (Sharon); “I think it’s definitely the biology of it [HD]” (Chris).

In terms of understanding how people adopted this perspective it seemed that this was due to the discourse around psychological difficulties in the context of HD provided by the health care professionals seen by participants. As Alice explained “they say they’re [psychological difficulties] part of the symptoms...they say that when you get to a further stage you’ll start to get a bit depressed”. As such, it seemed that this biological explanation of psychological distress was acceptable to people with HD. Interestingly, with regards to the explanations and information provided to people with HD, this seems understandable given

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that the healthcare professionals people had contact with were often medical professionals.

As such, participants were more frequently exposed to a medical understanding of psychological distress in HD.

Furthermore, one participant explained why they held a biological understanding of psychological distress: "It only makes sense when I think about it as part of the Huntington's biological thing" (Sharon). It seemed that this participant could only understand her emotional experience from a biological perspective, as to her, nothing else in her life could be responsible for it. Consequently, an explanation which removed the control from her over her emotions was more acceptable as it seemed that if she had control over how she felt then she would have already actively sought to change them. However, while this biological attributional process seemed to be participants' predominant experience there were also some additional views.

In addition to a biological understanding, some participants also commented that they were not confident as to the cause. There was an ambivalence which seemed to be driven by a feeling that health care professionals did not fully know the cause of psychological distress in HD. Lyndsey's experience was that "they often say they don't know if depression's linked to HD and that they don't know either way". It seemed that while participants were often accepting of a biological understanding there remained some confusion and uncertainty with Lyndsey going on to say "I said earlier I think it's just the HD but I don't. I think it's both [due to biological and psychological factors]". Therefore, it seemed that being given the space to reflect on their understanding and where this had originated, enabled acknowledgement of the potential for psychological difficulties to occur in response to living with HD. However, this psychological understanding seemed to be more implicit and subtle resulting from a belief that a biological explanation could not solely explain psychological

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distress in HD. Consequently, it appeared that participants were amenable to the two explanations existing in parallel.

Many participants seemed to experience HD as an uncertain and ambiguous condition which indirectly influenced the interpretations people made about their behaviour: "now everything's illness and it doesn't matter. You can't get that out of your head really" (Chris). Indeed, this uncertainty and ambiguity seemed to induce feelings of anxiety and/or low mood for many participants. As such, many participants reported having undertaken genetic testing to reduce the anxiety of not knowing whether they were gene carriers: "If I didn't have the test, I would feel anxious" (Sue) and "...I think I'd have been probably down and upset about it if I hadn't have had the test and just sat in limbo not knowing" (Anna). However confirmation of the gene seemed to result in further ambiguity with regards to living with HD. This dilemma was highlighted by one participant's experience: "it is that blessing and a curse to get to know that something massive is gonna happen to you that's not necessarily going to be pleasant" (Sue). Furthermore, participants explained how they could not be certain how and when they would be affected by the condition. This ambiguity was often described to lead to feelings of anxiety and low mood.

No one can tell you what kind of symptoms you're going to get. I suppose that makes you a bit anxious because you don't know ... And it's like, very hard to, you'll never know definitely even at the time when stuff happens, it's like he [doctor] said it can be any way kind of thing, it's not a set path which is really hard. (Alice)

While psychological distress was not attributed to the biological nature of HD, nonetheless, it was a result of living with HD. Additionally, despite not knowing exactly what may occur for people in the future, feelings of anxiety and low mood were

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also triggered by an understanding that the experience of physical and psychological difficulties was inevitable;

You know what's coming because you've seen it ... you just worry about, obviously you know what's going to happen so that makes you feel a bit depressed when you're thinking about it and a bit anxious that you know what's happening. (Alice)

I suppose it does feel like a ticking time bomb but it's meant to go on for quite a long time. (Lyndsey)

It appeared as though both the ambiguity and inevitability of the potential difficulties people could experience felt daunting. The anticipation of both physical and psychological difficulties seemed to induce feelings of anxiety and helplessness resulting in low mood for some participants. Additionally, many participants talked about their experience of psychological distress, in particular anxiety, in the context of worrying about the genetic transmission of HD.

You don't really want to think about that type of thing because at the moment I'm quite selfish. I just think about myself and get on with my day. It's almost like you can't cope with thinking about if the boys had it as well...but I always worry about the boys getting it. (Chris)

For those participants with children, this seemed to provide an additional cause of distress. It seemed that in order to manage this cause of distress avoidance was often used due to an inability to control the situation.

Indeed, all participants described their experience of HD as removing control from them which seemed to result in feelings of helplessness. One participant explained "...it [HD] takes over at the end of the day, I can't really do anything about it" (James). There was

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almost a sense of being resigned to the idea that people were in the hands of HD and had to accept “that lack of control in your life that, really, you’re not master of your own destiny at all despite what you might think” (Sue). This sense of helplessness was further highlighted by Chris: “sometimes you just feel like you’re living on a sinking ship”. It appeared that participants were very aware of the fact that there was currently no treatment or cure for HD and as such there was nothing they could do, resulting in feeling out of control.

### **Changes across time: “*in the past you’d just get on with it*”**

Further to people’s current understanding of their psychological difficulties, it seemed that some participants’ experience of their difficulties had changed from prior to having the HD gene confirmed, reflecting a dynamic process. Now participants attributed any instance of psychological distress to HD “whereas in the past you’d just get on with it” (Chris) and often would not pay much attention to becoming irritable or anxious. In particular, when people started to experience difficulties following finding out about the HD gene, it seemed that they were increasingly likely to attribute them to the biological progressive nature of HD: “when something suddenly changes like that you think, you automatically think well the cause might be HD” (Simon). Chris further explained that “you’ve got something to blame it on now”, describing “if you’re tired it’s because of the gene, you know, if you get annoyed it’s because of the gene”. This understanding was also described by Dave who commented “the question is do I over analyse? If I weren’t thinking about it would my mind actually bother about it? A couple of years ago I wouldn’t even have thought about it I would have just brushed it off”. These comments reflect the idea that once people know they have the HD gene, any difficulties are viewed through this lens and are subsequently attributed to HD. However, prior to knowing about the gene they were likely to attribute their experience of psychological distress differently and in some cases, ignore it or minimise it.

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Another specific change in attributional process for some participants regarded the origin of emotional responses, and whether these were part of their personality or part of the HD. As James explained "I don't know if it's probably the early signs of Huntington's back then or if it's just part of me". This conflict of understanding was further suggested by Sue who talked about her experience of irritability and anger as "part of the condition" but went on to say "but I'm a defensive person so that's part of my personality". Again, it appeared that the additional lens of HD had provoked a re-assessment as to where experiences came from and how they could be understood.

Again, participants also expressed the view that their experiences could be due to an interplay of factors. Dave seemed to understand his psychological distress as being part of his personality, however with the potential for HD to accentuate these: "I don't think the HD's brought them on, I think I've had them anyway...it's just I've always had them and now they could get worse because of this". Lyndsey similarly recognised longstanding difficulties which had changed: "I've always been inclined to get a bit down but this is on a completely different level", which seemed to suggest that her experience of psychological distress was enhanced in the context of HD. These comments highlight the process of change in participants' understanding of their experience. While people previously understood their psychological distress to be a part of their personality, the knowledge of HD had altered their understanding. As such there seemed to be an understanding that HD had increased previous levels of anxiety and low mood. Furthermore, HD had not only influenced how participants understood their present and future psychological distress but it had also influenced how they viewed their past experience of psychological distress.

Conversely, one participant continued to see their experience of psychological distress prior to and following confirmation of the HD gene as separate. On discussing experiencing a period of depression prior to knowing about HD, Simon commented "I don't attribute any

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of that to HD related stuff, that was really to do with work pressures". This highlights a situational and psychological understanding of his experience.

There certainly seemed to be an understanding from some participants that HD had the ability to accentuate some of the negative aspects of a person's personality. Therefore, it seemed that while they acknowledged that some difficulties were almost an inherent part of their personality, they believed that the HD may heighten some of these personality factors. Interestingly, there was no mention of the impact of HD on more positive emotional responses people experienced in their lives. In addition, Simon understood personality to be a dynamic process in itself: "I mean everybody's personality is different and the problem is personality is changing". Seemingly, Simon understood personality as having the potential for change. It therefore seemed that it may be hard to conclude the cause of psychological distress on the basis that personality changes over time. Therefore, there appeared to be an understanding that difficulties over time may have numerous potential causes and that while HD progresses over time, so can other causes such as personality.

### **Therapy instils hope and fight: *"a light at the end of the tunnel"***

Most participants, while not having had any psychological therapy before, described seeing the opportunity to engage in MBCT as positive. Most participants were not sure what to expect but there seemed to be a hope that psychological therapy could support them to manage and control the psychological difficulties they experienced: "I think for me it feels that maybe there is a bit of a light at the end of the tunnel" (Lyndsey).

It also seemed that people felt that taking part in therapy was a means of fighting against some of the difficulties HD could bring. This fight was articulated by Sharon who commented: "I wouldn't want to just be putting up with it if there was something I could do

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about it", with psychological therapy providing the possibility of being able to play a role in this. Furthermore, Simon explained "dealing with anxiety is important because it's there constantly...yep control of anxiety and worry is important", again with the hope that psychological therapy could contribute to this.

Six participants were on medication to help manage psychological distress. However, it seemed that most participants preferred the idea of psychological therapy to medication. This preference seemed to originate from the idea of medication being chemicals placed into the body whereas psychological therapy, if helpful, could provide participants with an alternative or additional approach to medication that was less intrusive: "I'd rather something more natural than medication" (Alice). This was further emphasised by Simon who commented:

The drugs out there at the moment are probably quite crude and may suppress other things...So I think from my point of view anything you can, as it were, do naturally and do by going through a process of erm, of psychological awareness and you know exercises if you like and routines has to be a good thing.

It was apparent that, where possible and optional, people were engaging in the therapy with the hope that this could provide them with an alternative, or in some cases additional, approach to manage the psychological difficulties they currently experienced and potentially may experience in the future. All participants talked about how they did not have any particular expectations of psychological therapy, rather the idea of accessing psychological therapy provided them with hope that their level of psychological distress, either now or in the future, could be managed or reduced.

Despite the hope people had for psychological therapy to help them with the psychological difficulties they experienced, due to their biological understanding of

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psychological difficulties in HD, there seemed to be some uncertainty as to how it might help. Alice discussed how she was “not too sure” about how therapy could help “because apparently everyone takes the tablets so I don't know”. It seemed that people did not have much information or understanding of psychological approaches, particularly within the context of HD, and as such were not able to contemplate how it would be effective. However, despite this uncertainty, the hope for psychological therapy to be beneficial to participants was maintained: “even if it's minimal the difference it makes, it still is worth doing” (Sue).

There seemed to be an understanding that engaging in psychological therapy required a certain mind set in order for the therapy to be beneficial, potentially as a result of the uncertainty of how it may help. A number of participants used the term “open-minded” as a characteristic they felt important when taking part. Dave explained “I'm always willing to try new things” while James commented “I'm open-minded to it and see where it goes you know, see what happens”. In part it seemed that this open-mindedness was required due to information people had received regarding the biological nature of some psychological difficulties:

I'm hoping I've got an open mind about it...because like I said we're all kind of, we're told you know that things are a certain way and that's you know kind of what we have to deal with like you know, low mood and depression etcetera.  
(Sharon)

Seemingly, while participants were hopeful that psychological therapy could help them manage any psychological difficulties they experienced, there was an element of reservation with regards to how much it could help. This was reflected in comments from some participants who expressed an understanding and expectation that psychological therapy

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would have its limits, particularly as HD progressed: “there’s probably a limit to how far it will go when it starts, you know, getting progressively worse...there’s probably a limit to what it can do” (Chris).

In addition to being open-minded, there also seemed to be an understanding that therapy would require effort on the part of the participant. The majority of participants seemed committed to actively engaging with the therapy. Sue commented “I think I’ve got to really make the effort” and Sharon explained “I’m going to do my best”. These comments regarding therapy requiring effort, and the concept of being open-minded, potentially reflected the dissonance between understanding psychological distress as a consequence of the biological neurodegenerative process of HD and adopting a psychological approach in managing this. When questioned regarding how participants thought a psychological approach could help considering many adopted a biological understanding, participants generally struggled to provide an answer: “I haven’t a clue. That’s what I’m I’m a little bit confused about, a lot confused about” (Sharon). However, despite this, it seemed that the hope that it could help people to manage their distress was more important to participants.

### **Discussion**

The current analysis of people with HD’s experience and understanding of psychological difficulties in HD and expectations of psychological therapy revealed three superordinate themes. Findings suggest that their understandings of the causes of psychological difficulties are varied with participants describing different potential causes of their psychological difficulties including both biological and psychological accounts. There was an acknowledgement that psychological difficulties were sometimes reactive in terms of being a response to living with HD. However, there was also a more dominant understanding

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that ran alongside a psychological account whereby psychological difficulties were attributed to the biological process of HD and, subsequently, were inevitable.

Indeed, psychological difficulties in HD are likely due to a combination of psychological and neurobiological factors (Weintraub & Burn, 2011). However, research has tended to emphasise neurobiological factors (e.g. Gregory et al., 2015; Van den Stock et al., 2015) above the more psychological explanations (e.g. Nimmagadda et al., 2011). These findings indicate that medical models are incorporated far more than psychological models in HD insofar as accounts to which people are exposed. This is consistent with the research looking at psychological difficulties in HD which have focussed on the biological causes (Gregory et al., 2015; Van den Stock et al., 2015).

Interestingly, understandings and experiences of psychological distress seemed to reflect a dynamic process for many participants as opposed to being static. In their self-regulation model of chronic illness, Leventhal, Meyer and Nernez (1980) propose that people, based on their experience of their illness, develop their own illness beliefs to help them make sense of their illness and subsequently cope with and adapt to their illness. Indeed it has been suggested that people's beliefs about their illness are often influenced, unsurprisingly, by the information they are surrounded by and as such these beliefs are changing dependent on the information to which a person is exposed (Leventhal, Leventhal & Cameron, 2001).

Furthermore, prior to the individual themselves finding out they have the gene, for many there was an awareness that HD was in the family and they were at risk. However, finding out they themselves had the gene resulted in an increased level of distress and a different understanding regarding their experience of distress, with most psychological distress now being attributed to HD. Therefore, the current research demonstrates how

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people's experience of psychological distress, across the progression of HD is likely to change and as such may require different approaches dependent on where the individual is in their HD journey.

In a systematic review examining the psychological impact of predictive testing, an initial increase in feelings of hopelessness was found (Crozier, Robertson & Dale, 2015). The current research also identified the hopelessness that some participants felt as a result of living with HD which further seemed to impact on their mood. Furthermore, how an individual perceives their chronic illness, including the sense of control, has been shown to contribute to both their physical and psychological well-being (Arran et al., 2013; Heijmans, 1998; Simpson, Lekwuwa & Crawford, 2013). Indeed, the hopelessness some participants felt seemed to be associated with participants' sense of control over their health and life in general. Certainly, it was apparent in the current research that many participants felt HD had taken this away.

In addition, the current findings support those of Arran et al. (2013) who found that people with HD felt they had little control, both personally and with regards to the treatment of HD. Indeed, in the current research, the option to engage in psychological therapy, in particular MBCT, appeared to enable participants to feel they were regaining some of the control they had lost and they hoped would enable them to feel more in control in the future. Similarly, they felt they were being proactive in improving their well-being as opposed to waiting for what they felt was inevitable. Thus increasing a person's perception of control over their illness may result in improved wellbeing (Hagger & Orbell, 2003).

However, similar to the experience of people with PD (Eccles, Murray & Simpson, 2011), due to the progressive degenerative nature of HD, it is unlikely, and potentially unrealistic, that people with HD will hold positive control beliefs. Consequently, it may be

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more effective to work with people with HD to accept and learn to live with reduced control and the ambiguity HD brings. Certainly, living with the unpredictable and uncontrollable nature of HD, acceptance is of particular importance (Helder et al., 2002). In fact, individuals undertaking MBCT, the therapy in which the participants were due to engage have emphasised its value in enabling acceptance (Mason & Hargreaves, 2001).

Further to struggling with the perception of a loss of control, the uncertainty associated with HD often resulted in feelings of anxiety. Indeed, anxiety has been shown to be one of the most common psychosocial responses to living with a chronic illness (Livneh & Antonak, 2005). Novak and Tabrizi (2010) noted that people can often find knowing they have the HD gene easier than the uncertainty of HD. While this was true for the majority of participants, this then resulted in a different uncertainty that people had to manage i.e. the uncertainty regarding when the disease would begin to affect them. The Huntington's Disease Society of America note "There's no typical person with HD. Each individual has complex unique needs" (1999, p.7). As such the unique and unpredictable nature of HD is likely to increase a person's anxiety, leaving them uncertain regarding their future and the impact the disease may have.

Additionally, due to the mean age of onset of around 40 years of age, gene carriers may have already passed the gene on to their children (Duisterhof, Trijsburg, Niermjer, Roos & Tibben, 2001). Subsequently, there were wider implications of having the HD gene than just those of the individual. Indeed for those who talked about having children, anxieties were discussed as a result of the potential to have passed the gene on. Furthermore, while each individual's experience of HD is likely to be different, most people, given its genetic transmission will have seen a family member, most likely a parent, develop the disease and will be familiar with the changes this causes (Novak & Tabrizi, 2010). Consequently, having seen the disease progress in a loved one and anticipating what their own disease progression

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may be, anxiety levels may be expectedly high. These were experiences described by many participants, by which it was apparent that a psychological understanding of distress was also accepted.

Furthermore, with regards to people's expectations of psychological therapy, participants felt it provided them with an alternative approach to medication, giving them hope of being able to manage and gain some control over their psychological difficulties believed to be associated with HD. Indeed, a number of participants currently taking medication commented on the hope that they may not have to take them in the long term. However, an important caveat is that all participants had already signed up to engage in a pilot trial of MBCT and as such may have already been open to psychological approaches and interventions. Consequently, there is the potential for a bias toward a psychological approach to have been reported as those who did not opt in to engage in the MBCT programme were not recruited.

Interestingly, previous studies have shown patient outcome expectations to be important in engagement and completion of therapy programmes, including CBT and MBCT (Snippe et al., 2015). "Outcome expectations reflect patients' prognostic beliefs about the consequences of engaging in treatment" (Constantino, Arnkoff, Glass, Ametrano & Smith, 2011, p.184). In a sample of people with diabetes, Snippe et al. (2015) found that people were more likely to complete and benefit from CBT and MBCT if they had high expectations of the outcomes.

In addition, context has been suggested to be a potential influence on an individual's expectations insofar as if a person has prior experience of a psychological therapy then it is likely that their expectations of future therapy will be influenced by their previous experience (Constantino et al., 2011). For example, a person who has had a positive experience of

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therapy is more likely to have higher expectations of future therapy than someone who had a negative experience of therapy. Of the participants in the current study, four had previously accessed therapy unrelated to HD, some of whom had a positive experience and some who did not find it helpful. However, this was often accompanied by the understanding that the timing of the therapy influenced how helpful it was. Indeed, despite some participants having a negative experience of previous therapy, this did not seem to influence their expectations of the MBCT course, potentially due to their hope that it would help.

Indeed, while expectations may be important in terms of engagement, the hope that is created as a result of patients' positive expectations may also influence outcomes (Frank, 1973). In the current study the concept of hope was discussed, in some cases explicitly and in others implicitly. While participants did not seem to have many expectations of psychological therapy it was apparent that, to many, it offered hope of being able to manage their psychological distress better.

Interestingly, despite a dominant biological understanding of distress, participants were interested in a psychological approach to its treatment, suggesting a certain level of dissonance. The theory of cognitive dissonance (Festinger, 1957) suggests that individuals have a tendency to seek consistency regarding their cognitions (i.e. beliefs). When there is not consistency, dissonance occurs. Participants were engaging in psychological therapy despite holding a biological understanding of psychological distress. However, it is suggested that there are many situations where dissonance is unavoidable (Festinger, 1962). Considering that there is no cure for HD and the desire of some participants to avoid medication where possible, this dissonance may have been compensated for with hope. Consequently, it may be suggested that even though dissonance can occur between a person's beliefs and their actions (i.e. holding a biological understanding and accessing psychological

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therapy), this is tolerable when the potential benefit of the dissonant action has the potential to outweigh this conflict.

### **Study Limitations**

However, this study has a number of limitations. The only individuals invited to be participants were those who had already consented to take part in an MBCT trial. People who had not signed up to the MBCT trial were therefore excluded. Subsequently, it may be suggested that these individuals may be more psychologically minded and more open to psychological approaches than those who declined to take part in the trial. Furthermore, the consideration of a psychological understanding of distress, on some occasions, seemed to be a result of taking part in the current research. It seemed that having time to think about an alternative perspective enabled people to reflect on their experience and understanding, something which may not have been the case otherwise. Therefore, it cannot be concluded that the understandings and experiences of psychological difficulties and expectations of psychological therapy, as described by participants, would reflect those of people who were not signed up to engage in MBCT. Consequently, it may be of interest to investigate the understanding and experience of psychological distress in people who were not open to the idea of engaging in psychological therapy.

Additionally, the lead researcher took part in the MBCT trial alongside participants in the current study (after having interviewed them). This took place prior to the completion of the data analysis. It is therefore possible that the lead researcher developed a greater insight into the lives and experience of participants than would have been possible during a 60-minute interview. However, in order to manage this, a reflective diary was kept to maintain an awareness of understanding that was obtained during the MBCT course compared with that obtained from the interviews.

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### **Clinical implications and future research**

The current research highlights a number of clinical implications. Firstly, it has been demonstrated that people with HD may be open to a psychological understanding of distress in HD and subsequently psychological approaches. Indeed, the openness to considering psychological factors as influential in the experience of distress and acceptance of a psychological approach has already been demonstrated in people with another neurodegenerative condition, PD (Oehlberg et al., 2008). Consequently, the provision of, and access to, psychological therapy services for people with HD should be considered. Indeed, findings (Tabrizi et al., 2012; Eidelberg & Surmeier, 2011) support the argument for non-pharmacologic approaches such as CBT for the management of behavioural difficulties such as irritability, either alongside or as an alternative to medication. Currently, psychological support is not prioritised in HD, potentially due to the understanding that psychological distress occurs as part of the HD process. However, here there is indication that psychological approaches may be acceptable to people with HD with the potential to improve well-being.

As the current research only examined the perspectives of people with pre-symptomatic HD it would be valuable, where possible, to obtain the perspectives of people at different stages of the HD process. It is possible that people with more advanced HD may struggle to engage with psychological therapy, particularly if there has been a significant impact on a person's cognition.

### **Conclusions**

Overall, the current research has demonstrated that participants accepted both a biological and psychological understanding of psychological distress, however with a biological view seeming to dominate. Furthermore, participants' experiences were

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changeable over time, dependent on the context in which the individual was experiencing distress. Finally, psychological therapy was accepted as an approach to support people to manage their distress. This was often accompanied with the hope this could provide an alternative or additional approach to medication that could support people with HD to feel more in control over their experience.

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**References**

- Arran, N., Craufurd, D. & Simpson, J. (2013). Illness perceptions, coping styles and psychological distress in adults with Huntington's disease. *Psychology, Health & Medicine, 19*, 169-179. doi: 10.1080/13548506.2013.802355.
- Aubeeluck, A., & Wilson, E. (2008). Huntington's disease. Part 1: essential background and management. *British Journal of Nursing, 17* (3), 146-151.
- Bury, M. (1982). Chronic illness as biographical disruption. *Sociology of health and illness, 4* (2), 167-182.
- Charidimou, A., Seamons, J., Selai, C., & Schrag, A. (2011). The role of Cognitive-Behavioural Therapy for patients with depression in Parkinson's disease. *Parkinson's disease, 2011*. doi: 10.4061/2011/737523.
- Constantino, M. J., Arnkoff, D. B., Glass, C. R., Ametrano, R. M., & Smith, J. Z. (2011). Expectations. *Journal of Clinical Psychology, 67* (2), 184–192. doi: 10.1002/jclp.20754.
- Craufurd, D., & Snowden, J.S. (2011). Neuropsychiatry and Neuropsychology. In G.P. Bates, S.J. Tabrizi, & L. Jones, *Huntington's Disease* (4<sup>th</sup> Edition). Oxford University Press, New York: NY.
- Craufurd, D., Thompson, J.C., & Snowden, J.S. (2001). Behavioral changes in Huntington's disease. *Neuropsychiatry, Neuropsychology and Behavioral Neurology, 14* (4), 219-226.
- Croizer, S., Robertson, N., & Dale, M. (2015). The psychological impact of predictive genetic testing for Huntington's disease: A systematic review of the literature. *Journal of Genetic Counselling, 24*, 29-39. doi: 10.1007/s10897-014-9755-y.

## PSYCHOLOGICAL DISTRESS IN HUNTINGTON'S DISEASE

- Dale, M., & van Duijn, E. (2015). Anxiety in Huntington's disease. *The Journal of Neuropsychiatry and Clinical Neurosciences*, 27 (4), 262-271. doi: 10.1176/appi.neuropsych.14100265.
- Dewhurst, K., Oliver, J., Trick, K.L.K., & McKnight, A L. (1969). Neuro-psychiatric aspects of Huntington's disease. *Confin Neurol*, 31 (4), 258-268. doi: 10.1159/000103486.
- Dobkin, R.D., Menza, M., Allen, L.A., Gara, M.A., Mark, M.H., Tiu, J., ... Friedman, J. (2011). Cognitive behavioural therapy for depression in Parkinson's disease: A randomised, controlled trial. *American Journal of Psychiatry*, 168 (10), 1066-1074. doi: 10.1176/appi.ajp.2011.10111669.
- Duff, K., Paulsen, J. S., Beglinger, L. J., Langbehn, D. R., Stout, J. C., & Predict-HD Investigators of the Huntington Study Group. (2007). Psychiatric symptoms in Huntington's disease before diagnosis: the predict-HD study. *Biological Psychiatry*, 62 (12), 1341-1346. doi: 10.1016/j.biopsych.2006.11.034.
- Duisterhof, M., Trijsburg, R.W., Niermeijer, M.F., Roos, R.A.C., & Tibben, A. (2001). Psychological studies in Huntington's disease: Making up the balance. *Journal of Medical Genetics*, 38, 852-861. doi: 10.1136/jmg.38.12.852.
- Eccles, F.J., Murray, C., & Simpson, J. (2011). Perceptions of cause and control in people with Parkinson's disease. *Disability and Rehabilitation*, 33, 1409-1420.
- Eidelberg, D., & Surmeier, D. J. (2011). Brain networks in Huntington disease. *The Journal of clinical investigation*, 121 (2), 484-492. doi: 10.1172/JCI45646.
- Festinger, L. (1957). *A theory of cognitive dissonance*. Stanford University Press.
- Festinger, L. (1962). *A theory of cognitive dissonance*. (Vol. 2). Stanford University Press.
- Fitzpatrick, L., Simpson, J. & Smith, A. (2010). A qualitative analysis of mindfulness-based cognitive therapy (MBCT) in Parkinson's disease. *Psychology and Psychotherapy: Theory, Research and Practice*, 83 (2), 179-192. doi: 10.1348/147608309X471514.

## PSYCHOLOGICAL DISTRESS IN HUNTINGTON'S DISEASE

- Frank, J. D. (1973). *Persuasion and healing* (2nd ed.). Baltimore: Johns Hopkins University Press.
- Gregory, S., Scahill, R.I., Seunarine, K.K., Stopford, C., Zhang, H., Zhang, J., ... & Track-HD Investigators. (2015). Neuropsychiatry and white matter microstructure in Huntington's disease. *Journal of Huntington's Disease*, 4, 239-249. doi: 10.3233/JHD-150160.
- Gu, J., Strauss, C., Bond, R., & Cavanagh, K. (2015). How do mindfulness-based cognitive therapy and mindfulness-based stress reduction improve mental health and well-being? A systematic review and meta-analysis of mediation studies. *Clinical Psychology Review*, 37, 1-12. doi: 10.1016/j.cpr.2015.01.006.
- Hagger, M.S., & Orbell, S. (2003). A meta-analytic review of the common-sense of illness representations. *Psychology and Health*, 18 (2), 141-184. doi: 10.1080/088704403100081321.
- Harper, D. (2011). Choosing a qualitative research method. In D. Harper & A.R. Thompson (Eds.) *Qualitative research methods in mental health and psychotherapy: A guide for students and practitioners*. John Wiley & Sons Ltd, p.83-98.
- Heijmans, M. (1998). Coping and adaptive outcome in chronic fatigue syndrome: Importance of illness cognitions. *Journal of Psychosomatic Research*, 45, 39-51.
- Helder, D.I., Kaptein, A.A., van Kempen, G.M.J., Weinman, J., van Houwelingen, H.C., & Roos, R.A.C. (2002). Living with Huntington's disease: Illness perceptions, coping mechanisms, and patients' well-being. *British Journal of Health Psychology*, 7 (4), 449-462. doi: 10.1348/135910702320645417.
- Ho, A.K., Gilbert, A.S., Mason, S.L., Goodman, A.O., & Barker, R.A. (2009). Health related quality of life in HD: Which factors matter most? *Movement Disorders*, 24 (4), 574-578. doi: 10.1002/mds.22412.

## PSYCHOLOGICAL DISTRESS IN HUNTINGTON'S DISEASE

Hoffman, N. (1999). Understanding the neuropsychiatric symptoms of Huntington's disease.

*The Journal of Neuroscience Nursing*, 31 (5), 309-313.

Huntington's Disease Society of America. (1999). *A Caregiver's Handbook for Advanced*

*Stage Huntington's disease*. Huntington's Disease Society of America, New York.

Julien, C.L., Thompson, J.C., Wild, S., Yardumian, P., Snowden, J.S., Turner, G. &

Craufurd, D. (2007). Psychiatric disorders in preclinical Huntington's disease.

*Journal of Neurology, Neurosurgery and Psychiatry*, 78, 939-943. doi:

10.1136/jnnp/2006.103309.

Kaptein, A. A., Helder, D. I., Scharloo, M., Van Kempen, G. M. J., Weinman, J., Van

Houwelingen, H. J. C. & Roos, R. a. C. (2006). Illness perceptions and coping explain

well-being in patients with Huntington's disease. *Psychology & Health*, 21 (4), 431-

446. doi: 10.1080/14768320500456947.

Kay, C., Fisher, E. & Hayden, M. R. (2014). Epidemiology. In Bates, G., Tabrizi, S. J. &

Jones, L. (eds.) *Huntington's disease*. 4th ed. Oxford: Oxford University Press.

Keenan, K.F., Simpson, S.A., Miedzybrodzka, Z., Alexander, D.A., & Semper, J. (2013).

How do partners find out about the risk of Huntington's disease in couple

relationships. *Journal of Genetic Counselling*, 22, 336-344. doi: 10.1007/s10897-012-

9562-2.

Kingma, E.M., van Duijn, E., Tinman, R., van der Mast, R.C., Raymund, A.C., & Ross, M.D.

(2008). Behavioural problems in Huntington's disease using the Problem Behaviours

Assessment. *General Hospital Psychiatry*, 30, 155-161. doi:

10.1016/j.genhosppsy.2007.11.005.

## PSYCHOLOGICAL DISTRESS IN HUNTINGTON'S DISEASE

- Kirkwood, S.C., Su, J.L., Conneally, M., & Foroud, T. (2001). Progression of symptoms in the early and middle stages of Huntington's disease. *Archives of Neurology*, 58 (2), 273-278. doi: 10.1001/archneur.58.2.273.
- Kloppel, S., Stonnington, C.M., Petrovic, P., Mobbs, D., Tuscher, O., Craufurd, D., ... Frackowiak, R.S.J. (2010). Irritability in pre-clinical Huntington's disease. *Neuropsychologia*, 48, 549-557. doi: 10.1016/j.neuropsychologia.2009.10.016.
- Kowalski, P.C., Belcher, D.C., Keltner, N.O., Dowben, J.S. (2015). Biological Perspectives: Huntington's disease. *Perspectives in psychiatric care*, 51 (3), 157-161. doi: 10.1111/ppc.12121.
- Kremer, B. (2002). Clinical neurology in Huntington's disease. In: Bates, G., Harper, P. & Jones, L. (eds.) *Huntington's disease*. 3rd ed. Oxford: Oxford University Press.
- Leventhal, H., Leventhal, E., & Cameron, L. (2001). Representations, procedures, and affect in illness self-regulation: A perceptual-cognitive model. In Baum, A., Revenson, T., Singer, J. (eds). *Handbook of health psychology*. Hillsdale, NJ: Lawrence Erlbaum Associates (p.19-47).
- Leventhal, H., Meyer, D., & Nerenz, D. (1980). The common sense representation of illness danger. *Contributions to medical psychology*, 2, 7-30.
- Livneh, H. & Antonak, R.F. (2005). Psychosocial adaptation to chronic illness and disability: A primer for counsellors. *Journal of counselling and development*, 83 (1), 12-20. doi: 10.1002/j.1556-6678.2005.tb00575.x.
- Mason, O. & Hargreaves, I. (2001), A qualitative study of mindfulness-based cognitive therapy for depression. *British Journal of Medical Psychology*, 74, 197-212. doi: 10.1348/000711201160911.

## PSYCHOLOGICAL DISTRESS IN HUNTINGTON'S DISEASE

- Meiser, B., & Dunn, S. (2000). Psychological impact of genetic testing for Huntington's disease: An update of the literature. *Journal of Neurology, Neurosurgery and Psychiatry*, *69* (5), 574-578. doi: 10.1136/jnnp.69.5.574.
- Nimmagadda, S.R., Agrawal, N., Worrall-Davies, A., Markova, I., & Rickards, H. (2011). Determinants of irritability in Huntington's disease. *Acta Neuropsychiatrica*, *23*, 309-314. doi: 10.1111/j.1601-5215.2011.005653.x.
- Novak, M.J.U., & Tabrizi, S.J. (2010). Huntington's disease. *BMJ*, *340*, 34-40. doi: 10.1136/bmj.c3109.
- Oehlberg, K., Barg, E K., Brown, G. K., Taraborelli, D., Stern, M. B., & Weintraub, D. (2008). Attitudes regarding the etiology and treatment of depression in Parkinson's disease: A qualitative study. *Journal of Geriatric Psychiatry and Neurology*, *21*, 123-132. doi: 10.1348/147608309X471514.
- Paulsen, J.S., Nehl, C., Hoth, K.F., Kanz, J.E., Benjamin, M., Conybeare, R., ... & Turner, B. (2005). Depression and stages of Huntington's disease. *Journal of Neuropsychiatry and Clinical Neurosciences*, *17* (4), 496-502. doi: 10.1176/jnp.17.4.496.
- Pla, P., Orvoen, S., Saudou, F., David, D.J., & Humbert, S. (2014). Mood disorders in Huntington's disease: From behavior to cellular and molecular mechanisms. *Frontiers in Behavioral Neuroscience*, *8* (135), 1-15. doi: 10.3389/fnbeh.2014.00135.
- Segal, Z.V., Williams, J.M., & Teasdale, J. (2002). *Mindfulness-based cognitive therapy for depression: A new approach to preventing relapse*. London, UK: Guildford Press.
- Simpson, J., Lekwuwa, G., & Crawford, T. (2013). Illness beliefs and psychological outcome in people with Parkinson's disease. *Chronic Illness*, *9* (2), 165-176. doi: 10.1177/1742395313478219.

## PSYCHOLOGICAL DISTRESS IN HUNTINGTON'S DISEASE

- Smith, J.A., Flowers, P. & Larkin, M. (2009). *Interpretative phenomenological analysis: theory method and research: understanding method and application*. London, Sage.
- Smith, J.A., & Osborn, M. (2003). Interpretative Phenomenological Analysis. In J.A. Smith (Ed.), *Qualitative Psychology*. London: Sage.
- Snippe, E., Schroevers, M.J., Tovote, K.A., Sanderman, A., Emmelkamp, P.M.G., & Fleer, J. (2015). Patients' outcome expectations matter in psychological interventions for patients with diabetes and comorbid depressive symptoms. *Cognitive Therapy Research, 39*, 307-317. doi: 10.1007/s10608-014-9667-z.
- Tabrizi, S. J., Reilmann, R., Roos, R. A., Durr, A., Leavitt, B., Owen, G., ... & Kennard, C. (2012). Potential endpoints for clinical trials in premanifest and early Huntington's disease in the TRACK-HD study: Analysis of 24 month observational data. *The Lancet Neurology, 11*(1), 42-53. doi: 10.1016/S1474-4422(11)70263-0.
- Van den Stock, J., De Winter, F-L., Ahmad, R., Sunaert, S., Van Laere, K., Vandenberghe, W., & Vandenberghe, M. (2015). Functional brain changes underlying irritability in premanifest Huntington's disease. *Human Brain Mapping, 36*, 2681-2690. doi: 10.1002/hbm.22799.
- Van Duijn, E., Kingma, E.M., & van der Mast, R.C. (2007). Psychopathology in verified Huntington's disease gene carriers. *Journal of Neuropsychiatry and Clinical Neuroscience, 19* (4), 441-448. doi: 10.1176/appi.neuropsych.19.4.441.
- Weintraub, D., & Burn, D. J. (2011). Parkinson's disease: The quintessential neuropsychiatric disorder. *Movement Disorders, 26* (6), 1022-1031. doi: 10.1002/mds.23664.
- Yardley, L. (2000). Dilemmas in qualitative health research. *Psychology and Health, 15* (2), 215-228. doi: 10.1080/08870440008400302.

## **Appendix 2-A: Background and methods of the Mindfulness-based cognitive therapy trial**

There has been little development regarding psychological interventions for people with Huntington's disease (HD). However, they are being developed for people with different neurological conditions such as Parkinson's disease (PD; Dobkin et al., 2011). Mindfulness-based cognitive therapy (MBCT) has previously been piloted for people with PD who reported an improvement in psychological wellbeing (Fitzpatrick et al., 2010). Consequently, a pilot trial of MBCT is being run for people with HD to see whether it would be an acceptable and useful approach for people with HD.

Additionally, due to the psychological and emotional consequences of HD, it also affects the people with whom they live (Aubeeluck et al., 2012). For example, caregiver depression has been shown to be associated with depression in the person with HD (Banaszkiewicz et al., 2012). Therefore the study will also obtain the views of a family member of the person with HD.

Both qualitative (semi-structured interviews) and quantitative (e.g. Hospital Anxiety and Depression Scale and the Five Facet Mindfulness Questionnaire) data were collected from both the person with HD and their family member. Data were collected both pre and post intervention.

To be included in the study participants had had the genetic test confirming the CAG expansion on the huntingtin gene and were all pre-symptomatic. All participants had clinical signs of low mood or depression identified in their notes or information recorded at their last clinic visit. Participants were aged 18 years or over and had not had changes in their medication six weeks prior to the start of the MBCT intervention. Participants were excluded

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if they currently had suicidal intent. Once the person with HD had been recruited they were asked if they have a family member or close friend who wished to participate.

## Appendix 2-B

Table 1: Extract from Sharon's transcript with initial summary notes and emerging themes

Initial Notations	Extract	Emerging Themes
<p>Don't know how therapy can help</p> <p>Need to be open minded</p> <p>Hopeful that therapy can help</p>	<p>P: Well since erm, well obviously, well (nurse) was saying about this course, I mean I didn't realise completely what it was you know, and then the little bit of reading I've done since and everything and I'm thinking okay, but I honestly don't know I'm just going to keep an open mind about it but im hopeful that just, you know, that I'll be feeling a little bit better, will make me you know more confident to go out and do things then that's good.</p> <p>I: So given that erm, kind of your main understanding of its psychological aspects such as mood and anxiety are cause by the Huntington's how do you think, so for example say it's got a biological cause, how do you think a psychological therapy can...</p>	<p>Open-minded</p> <p>Hope</p>
<p>Confusion about how therapy can help</p> <p>Trying to be open-minded</p> <p>A bit of help is better than nothing - hope</p>	<p>P: I haven't a clue, that's what I'm a little bit confused about, a lot confused about, but that why I'm trying to keep an open mind about the course and you know, and it might help research in the future so I don't know if it will but if it clears my mind then it will never work but it might just a bit and that's better than nothing when you don't have a lot of hope or anything when it gets to this point, so I don't know. Those are the questions I've been asking myself, they really are so that's why I'm quite looking forward to when it starts. I'm not going to put like, huge you know, pressure on it, on myself as well but I'm going to do my best. You know, and I think as well because I've been taking part in some research and there have been some results form that as well now and I'm thinking right okay maybe, there may be with this as well, this research so I'm hoping. We don't know until we do it.</p>	<p>Confusion/uncertainty</p> <p>Open-minded</p> <p>Hope</p>
<p>Not going to pressure self but will need to try – effort</p> <p>Previous experience of research has had positive results</p>	<p>P: I haven't a clue, that's what I'm a little bit confused about, a lot confused about, but that why I'm trying to keep an open mind about the course and you know, and it might help research in the future so I don't know if it will but if it clears my mind then it will never work but it might just a bit and that's better than nothing when you don't have a lot of hope or anything when it gets to this point, so I don't know. Those are the questions I've been asking myself, they really are so that's why I'm quite looking forward to when it starts. I'm not going to put like, huge you know, pressure on it, on myself as well but I'm going to do my best. You know, and I think as well because I've been taking part in some research and there have been some results form that as well now and I'm thinking right okay maybe, there may be with this as well, this research so I'm hoping. We don't know until we do it.</p>	<p>Active role in therapy</p> <p>Hope</p>

## Appendix 2-C: Journal Author Guidelines

### Submission

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The main emphasis of *Health Psychology*<sup>®</sup> is on original research in health psychology. Analytical reviews of research and brief scientific reports are also considered for publication. Submissions are welcomed from authors in psychology and other health-related disciplines.

Submit manuscripts electronically (.rtf, PDF, or .doc) to

Anne E. Kazak  
Center for Healthcare Delivery Science  
A.I. du Pont Hospital for Children  
Administration and Research Building, Room 281  
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Wilmington, DE 19803



[Manuscript Submission Portal Entrance](#)

Keep a copy of the manuscript to guard against loss. Do not submit manuscripts via mail or email.

In recognition of the reality that institutional spam filters may capture files from the APA and the Journals Back Office, please take the following steps to facilitate communication with our editorial office:

- Provide an alternative email address which we can use to contact you in the event of technical difficulties with email communication using your primary address;
- Add "apa.org" to your list of "safe" addresses and consider asking your IT administrators to add it to their "white list;" and
- Contact [Lindsay MacMurray](#) if you do not receive confirmation of your submission within three business days or an editorial decision letter within three months.

General correspondence may be directed to the [Editor's Office](#).

### Information About Submissions

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The page limit for research manuscripts is 25–30 pages. The page limit is inclusive of **all** parts of the manuscript, including the cover page, abstract, text, references, tables and figures.

Authors may request consideration of longer papers, in advance of submission, when there is clear justification for additional length (e.g., the paper reports on two or more studies or has an unusual or complex methodology).

Scholarly reviews and meta-analyses should not exceed 25 pages, but tables and references may be outside this page limit.

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Brief reports are encouraged for innovative work that may be premature for publication as a full research report because of small sample size, novel methodologies, etc. Brief reports should be designated as such and should not exceed a total of 12 pages, inclusive of **all** parts of the manuscript, including the cover page, abstract, text, references, tables and figures.

All manuscripts should be double-spaced, with margins of at least 1 inch on all sides and a standard font (e.g., Times New Roman) of 12 points (no smaller).

On the submission portal you will be asked to provide contact information for three individuals who are qualified to serve as unbiased reviewers for your paper. These people must have published peer reviewed work in a relevant field. They must be without any real or perceived conflict of interest with you and your co-authors. They cannot be at the same institution as any author, cannot be a co-author on any publications, and must not be a former or current trainee, advisor or mentor, etc.

*Health Psychology* considers letters concerning previously published articles. Letters should be no more than 500 words and have a maximum of five references.

Authors also have the option of placing supplemental materials online.

Submissions that exceed the page limits will be returned to the author for shortening prior to the initiation of peer review.

### Submission Letter

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The cover letter should indicate that the authors have read and followed the *Health Psychology* Instructions for Authors. It should also include a statement indicating that the paper has been seen and approved by all authors. The cover letter should describe how the paper advances research in health psychology, referring to the journal mission to assure that the submission fits with the types of papers published in *Health Psychology*.

The full mailing address, telephone, fax, and email address for the corresponding author should be included in the cover letter and title page, along with the names and affiliations of all co-authors.

The cover letter must confirm that the manuscript has not been published, is not currently submitted elsewhere, and that it does not contain data that is currently submitted or published elsewhere.

When a manuscript contains data that is part of a larger study, authors should describe the larger study and provide references for other study papers. Authors must be prepared to provide copies of related manuscripts when requested as part of the editorial review process. Authors should clarify the relationship between their paper, including detailed specification of the overlap in participants, measures, and analysis, and others from the study. The value-added scientific contribution of their study must be clearly stated in the cover letter.

Authors of brief reports should indicate in the cover letter that the full report is not under consideration for publication elsewhere and similarly address potential overlap with other papers.

### Manuscripts

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## PSYCHOLOGICAL DISTRESS IN HUNTINGTON'S DISEASE

The manuscript title should be accurate, fully explanatory, and no longer than 12 words. The title should reflect the content and population studied. If the paper reports a randomized clinical trial, this should be indicated in the title. The title of brief reports should start with the words "Brief Report".

The title page should include the names of all authors and their affiliations at the time the research was done. This information will be masked to ensure a blind peer review process by the editorial office. Authors should make sure that all other identifying information in the text of the paper is masked/removed prior to submission.

All manuscripts must include a structured abstract containing a maximum of 250 words with the following sections:

- Objective (brief statement of the purpose of the study);
- Methods (summary of the participants, design, measures, procedure);
- Results (primary findings); and
- Conclusions (specific statement of the implications of the data).

Please supply up to five keywords or brief phrases after the abstract. The Introduction should not exceed 3–4 pages in length. The paper should be referenced appropriately but excessive citations should be avoided.

All research involving human participants must describe oversight of the research process by the relevant Institutional Review Boards and should describe consent and assent procedures briefly in the Methods section.

All statistical tests should include effect size whenever possible.

First person language ("I", "we") should be avoided. Terminology should be sensitive to the individual who has a disease or disability. The journal endorses the concept of "people first, not their disability." Terminology should reflect the "person with a disability" (e.g., children with diabetes, persons with HIV infection, families of people with cancer) rather than the condition as an adjective (e.g., diabetic children, HIV patients, cancer families). Nonsexist language should be used.

It is important to highlight the significance and novel contribution of the work. The translation of research into practice must be evidenced in all manuscripts. Authors should incorporate a meaningful discussion of the clinical and/or policy implications of their work throughout the manuscript, rather than simply providing a separate section for this material.

*Health Psychology* publishes a broad array of types of papers. Authors of qualitative and measure development papers should read the guidelines for these types of papers, noted below.

### Qualitative Research

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Research papers that utilize qualitative methods should follow the general instructions to authors for style and format. We ask that authors of qualitative papers review the additional guidance below to assure that papers meet the following criteria utilized by *Health Psychology*.

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The introduction should make a compelling case for the significance of the study and clearly identify if the study is a stand-alone study or if it fits into a larger study. For example, qualitative manuscripts may inform the development of a survey, use small-incident samples, or establish feasibility. The specific qualitative paradigm should be specified (e.g., grounded theory, qualitative descriptive approach, interpretive phenomenology) with a rationale as to why it was selected to address the research question.

At the same time, authors are encouraged to avoid methodological tutorials and cite appropriate references for the methodology. Describe your sampling frame clearly and how the sample was selected, justifying the type and size of your sample using appropriate language for qualitative studies.

While many qualitative studies may not use a conceptual model, if you have done so, explain how the model may have shaped the design, data collection, analysis and interpretation. Explain carefully how you strengthened and insured rigor in your study e.g., data analysis protocols (including how coders were trained), audit procedures, and demonstration of data saturation. Describe the data analysis and how it relates to your overall approach or paradigm. Present rich and compelling results with data that have been analyzed and interpreted appropriately for your method (e.g., discourse analytic results would be presented differently than those of a grounded theory).

The paper should convey how this research fills an important gap in the science and promises to change the way we approach future studies.

### Scale Development

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Empirical papers related to the development of new instruments related to health psychology should follow the general guidelines for style and format of this journal. Authors should make a convincing case for the need and rationale for the new instrument, particularly with respect to new and innovative constructs. Included in this rationale should be the theoretical foundation on which their new instrument rests along with presentation of other, related scales currently in use.

It is important that the research have a degree of generalizability across populations and settings. Instruments that are more narrow in scope or of limited clinical utility may be better suited for subspecialty journals.

Authors should clearly articulate the specifics of the study design and of the analytical techniques used. There should be strong consistency among the purpose statements, methods, and the manner in which findings are presented.

An increasing number of studies are incorporating mixed-methods designs in their research. The specifics of these designs should be equally well-detailed without being excessive. Attention should be given to the nature of the items, the basis for their creation, and the rationale for the response options.

The underlying theoretical structure of the approach should be evident, for example, whether one is premising their study on classical or modern theory (IRT, Rasch) techniques. The characteristics of the research will be in part dictated by the nature of the scale. For instance, large, nationally-normed tests may have a much different make-up than that of small, more narrowly-defined measures. Research involving both types of instruments will be considered.

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Finally, all instrument development papers should convey how the literature base will be strengthened with the addition of the particular instrument along with a clear and convincing case for the clinical relevance of the information that it provides.

### Letters to the Editor

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*Health Psychology* will, at the discretion of the Editor-in-Chief, publish Letters to the Editor on the journal website.

Letters should be prepared in direct response to articles published in the journal, should include reference to the published paper in the letter, and should be sent to the [Editorial Manuscript Coordinator, Lindsay MacMurray](#) within 60 days of the date when the relevant article is published in hard copy.

The text of the letter, excluding the title, references and author(s) name, title, affiliation and email, may not exceed 400 words.

In a separate cover letter, the author should indicate that the submission is a Letter to the Editor for consideration of posting on the *Health Psychology* website and provide the full citation of the original article to which the letter refers. The cover letter should also indicate if the letter writer(s) have any conflicts of interest related to the article or correspondence.

Note: Letters will not be a forum for ongoing dialogue.

### Masked Review Policy

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Masked review is used. **Do not** include author information (addresses, phone numbers, electronic mail addresses, and fax numbers) in the manuscript.

Please ensure that the final version for production includes a byline and full author note for typesetting.

### Use of CONSORT Reporting Standards

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All randomized controlled trials must include a diagram indicating participant flow into the study and a completed CONSORT checklist. CONSORT diagrams (and adaptations) should be included whenever possible to clarify the flow of participants through a study.

### Manuscript Preparation

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Prepare manuscripts according to the [Publication Manual of the American Psychological Association \(6<sup>th</sup> edition\)](#). Manuscripts may be copyedited for bias-free language (see Chapter 3 of the *Publication Manual*).

Review APA's [Checklist for Manuscript Submission](#) before submitting your article.

Double-space all copy. Other formatting instructions, as well as instructions on preparing tables, figures, references, metrics, and abstracts, appear in the *Manual*. Additional guidance on APA Style is available on the [APA Style website](#).

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Below are additional instructions regarding the preparation of display equations, computer code, and tables.

### Display Equations

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We strongly encourage you to use MathType (third-party software) or Equation Editor 3.0 (built into pre-2007 versions of Word) to construct your equations, rather than the equation support that is built into Word 2007 and Word 2010. Equations composed with the built-in Word 2007/Word 2010 equation support are converted to low-resolution graphics when they enter the production process and must be rekeyed by the typesetter, which may introduce errors.

To construct your equations with MathType or Equation Editor 3.0:

- Go to the Text section of the Insert tab and select Object.
- Select MathType or Equation Editor 3.0 in the drop-down menu.

If you have an equation that has already been produced using Microsoft Word 2007 or 2010 and you have access to the full version of MathType 6.5 or later, you can convert this equation to MathType by clicking on MathType Insert Equation. Copy the equation from Microsoft Word and paste it into the MathType box. Verify that your equation is correct, click File, and then click Update. Your equation has now been inserted into your Word file as a MathType Equation.

Use Equation Editor 3.0 or MathType only for equations or for formulas that cannot be produced as Word text using the Times or Symbol font.

### Computer Code

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Because altering computer code in any way (e.g., indents, line spacing, line breaks, page breaks) during the typesetting process could alter its meaning, we treat computer code differently from the rest of your article in our production process. To that end, we request separate files for computer code.

#### **In Online Supplemental Material**

We request that runnable source code be included as supplemental material to the article. For more information, visit [Supplementing Your Article With Online Material](#).

#### **In the Text of the Article**

If you would like to include code in the text of your published manuscript, please submit a separate file with your code exactly as you want it to appear, using Courier New font with a type size of 8 points. We will make an image of each segment of code in your article that exceeds 40 characters in length. (Shorter snippets of code that appear in text will be typeset in Courier New and run in with the rest of the text.) If an appendix contains a mix of code and explanatory text, please submit a file that contains the entire appendix, with the code keyed in 8-point Courier New.

### Tables

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## PSYCHOLOGICAL DISTRESS IN HUNTINGTON'S DISEASE

Use Word's Insert Table function when you create tables. Using spaces or tabs in your table will create problems when the table is typeset and may result in errors.

### Submitting Supplemental Materials

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APA can place supplemental materials online, available via the published article in the PsycARTICLES® database. Please see [Supplementing Your Article With Online Material](#) for more details.

### References

---

List references in alphabetical order. Each listed reference should be cited in text, and each text citation should be listed in the References section.

Examples of basic reference formats:

- **Journal Article:**  
Hughes, G., Desantis, A., & Waszak, F. (2013). Mechanisms of intentional binding and sensory attenuation: The role of temporal prediction, temporal control, identity prediction, and motor prediction. *Psychological Bulletin*, 139, 133–151. <http://dx.doi.org/10.1037/a0028566>
- **Authored Book:**  
Rogers, T. T., & McClelland, J. L. (2004). *Semantic cognition: A parallel distributed processing approach*. Cambridge, MA: MIT Press.
- **Chapter in an Edited Book:**  
Gill, M. J., & Sypher, B. D. (2009). Workplace incivility and organizational trust. In P. Lutgen-Sandvik & B. D. Sypher (Eds.), *Destructive organizational communication: Processes, consequences, and constructive ways of organizing* (pp. 53–73). New York, NY: Taylor & Francis.

### Figures

---

Graphics files are welcome if supplied as Tiff or EPS files. Multipanel figures (i.e., figures with parts labeled a, b, c, d, etc.) should be assembled into one file.

The minimum line weight for line art is 0.5 point for optimal printing.

For more information about acceptable resolutions, fonts, sizing, and other figure issues, [please see the general guidelines](#).

When possible, please place symbol legends below the figure instead of to the side.

APA offers authors the option to publish their figures online in color without the costs associated with print publication of color figures.

The same caption will appear on both the online (color) and print (black and white) versions. To ensure that the figure can be understood in both formats, authors should add alternative wording (e.g., "the red (dark gray) bars represent") as needed.

## PSYCHOLOGICAL DISTRESS IN HUNTINGTON'S DISEASE

For authors who prefer their figures to be published in color both in print and online, original color figures can be printed in color at the editor's and publisher's discretion provided the author agrees to pay:

- \$900 for one figure
- An additional \$600 for the second figure
- An additional \$450 for each subsequent figure

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Authors of accepted papers must obtain and provide to the editor on final acceptance all necessary permissions to reproduce in print and electronic form any copyrighted work, including test materials (or portions thereof), photographs, and other graphic images (including those used as stimuli in experiments).

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### Ethical Principles

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It is a violation of APA Ethical Principles to publish "as original data, data that have been previously published" (Standard 8.13).

In addition, APA Ethical Principles specify that "after research results are published, psychologists do not withhold the data on which their conclusions are based from other competent professionals who seek to verify the substantive claims through reanalysis and who intend to use such data only for that purpose, provided that the

## PSYCHOLOGICAL DISTRESS IN HUNTINGTON'S DISEASE

confidentiality of the participants can be protected and unless legal rights concerning proprietary data preclude their release" (Standard 8.14).

APA expects authors to adhere to these standards. Specifically, APA expects authors to have their data available throughout the editorial review process and for at least 5 years after the date of publication.

Authors are required to state in writing that they have complied with APA ethical standards in the treatment of their sample, human or animal, or to describe the details of treatment.

- [Download Certification of Compliance With APA Ethical Principles Form \(PDF, 26KB\)](#)

The APA Ethics Office provides the full [Ethical Principles of Psychologists and Code of Conduct](#) electronically on its website in HTML, PDF, and Word format. You may also request a copy by [emailing](#) or calling the APA Ethics Office (202-336-5930). You may also read "Ethical Principles," December 1992, *American Psychologist*, Vol. 47, pp. 1597–1611.

**Section 3: Critical Appraisal**

**Understandings of psychological difficulties in people with Huntington's disease and their expectations of psychological therapy**

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Trainee Clinical Psychologist

Doctorate in Clinical Psychology

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Word Count: 3368

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## Critical Appraisal

Reflections on: Understandings of psychological difficulties in people with Huntington's disease and their expectations of psychological therapy'

This paper will provide a summary of the research findings and the study's strengths and limitations. It will also provide a reflective account of some of the process issues encountered throughout the research process, including the lead researcher attending the mindfulness-based cognitive therapy (MBCT) group and the impact of this on data analysis. It will also explore the current lack of psychological services for people with HD, reflecting on potential reasons for this and how it may be overcome in future.

### Research overview

The empirical paper was a qualitative exploration of people with HD's understanding of psychological distress and expectations of psychological therapy. Participants were all people with HD and were recruited from an existing therapy trial. Semi-structured interviews were used to understand participants' experiences and analysed using interpretative phenomenological analysis (IPA) following the steps described by Smith and Osborn (2003).

### Summary of research findings

The investigation of people with HD's understanding of psychological distress and expectations of psychological therapy resulted in the development of three superordinate themes: (1) Attributing psychological distress to HD: "*you're blaming everything on that now*"; (2) Changes across time: "*in the past you'd just get on with it*"; (3) Therapy instils hope and fight: "*a light at the end of the tunnel*".

The first theme reflected the understanding that HD was the cause of psychological distress, from both a biological and psychological perspective. Indeed, a biological

understanding appeared to be the dominant understanding people held. This seemed to come from information they were given by healthcare professionals that psychological difficulties are part of the progression of the disease. Additionally, there seemed to be some ambivalence as to the cause which appeared to result in a more implicit psychological understanding of distress. Similarly, the ambiguity and uncertainty associated with HD also seemed to be a contributing factor to participants' psychological distress. Consequently, whichever perspective participants held, and in some cases both perspectives were held simultaneously, the underlying cause was HD.

In addition to how participants attributed their psychological distress, the second theme reflected how this attribution appeared to reflect a dynamic process. This varied at different time points for example, prior to finding out about the HD gene, following receiving confirmation of the HD gene, and living with the anticipation of disease onset. Furthermore, not only did it change over time but the knowledge of having the HD gene influenced how participants perceived their previous experience. As such, participants' beliefs about their psychological distress altered from how they had previously viewed it, thus influencing perception from the past, present and future.

Finally, the third theme reflected the understanding that psychological therapy had the potential to give people with HD some control over their psychological experience. Indeed, it seemed to instil hope into participants and provide a sense that, while they could not control the motor aspects of the disease, they could fight against the psychological difficulties they experienced. Therefore, it enabled them to take an active role rather than remain passive, waiting for difficulties to occur. A number of participants described currently taking medication to manage the difficulties they experienced, however the option of a more natural approach appealed to many. However, alongside this hope was the understanding that

psychological therapy would require a certain mind-set and conscious effort on the part of the participant.

### **Strengths and Limitations**

A strength of the current research is that this is the first study, to my knowledge, to interview people with HD about their understanding of psychological distress and expectations of psychological therapy. It has therefore provided insight into the lived experience of people with HD, not previously sought.

There are, however, a number of limitations to the current study. The participants who took part had agreed to take part in an MBCT programme and were therefore potentially more open-minded to psychological approaches. This may not have had an impact on their understanding of psychological distress, but it may be that their willingness to engage in MBCT is reflective of their perspective of psychological approaches. Indeed, despite most participants having limited expectations of the course, no one commented that they thought it would be a waste of time. Furthermore, after I had collected the data, I took part in the MBCT group and therefore had more contact with participants than would have occurred otherwise and as such gained more insight into participants' experiences of psychological distress. Therefore, there was a potential for this to have influenced the data analysis process.

### **Reflections on the interview process**

Considering the research topic, investigating psychological distress and psychological therapy, it seems important to consider my potential influence on the interviews from the perspective of a trainee clinical psychologist. When conducting research interviews there is the potential for the researcher to influence participants' responses as a result of the interaction process and potential factors such as social desirability (Hewitt, 2007).

Consequently, coming from the position of a trainee clinical psychologist, and participants'

awareness of this, I felt it was important to remain aware of my understanding of psychological distress and the beliefs I hold about the benefits of psychological therapy in order to ensure these were not explicitly revealed during the interviews. I felt there could be a risk that coming from a psychological perspective would influence participants to want to talk about psychological distress and therapy from both a psychological and positive perspective. Consequently, I tried to ensure my follow-up questions and responses remained neutral and did not lead participants' responses. On reflection, I feel that participants described their experience that was true to them, particularly considering people described a biological understanding.

Furthermore, I found it difficult listening to participants' experiences of psychological distress, particularly hearing them describe the limited psychological support they had. Furthermore, I think this experience was made more difficult as a result of my attendance at the MBCT group. By having more contact with participants and getting to know more about them and their families, I felt a great sense of empathy for the situation they were in. I also felt inspired by the strength and resilience everyone showed in the face of HD and what this meant for their future. Additionally, this felt particularly difficult considering I was in the role of researcher, rather than a trainee clinical psychologist, which requires a different approach and the use of different skills. In my role as a trainee clinical psychologist I aim to understand a person's experience and work collaboratively to effect change. However, in the role as a researcher the aim is to obtain and understand a person's experience (Drury, Francis & Chapman, 2007) without working towards changing their experience.

Conversely, Wilde (1992) suggests that therapeutic skills can enhance the research process and in fact these skills cannot be completely put aside. Therefore, I felt able to use my clinical skills, such as active listening and empathy, to ensure participants felt heard and

understood while ensuring my own beliefs were not expressed. I feel this enabled participants to feel comfortable talking about their experience.

### **Considering the researcher's influence on data analysis**

Prior to conducting the research I did not have any prior experience of HD from either a research or clinical perspective. I had some previous research and clinical experience of working with another neurodegenerative disease, Parkinson's disease (PD). However due to the life-limiting nature of HD this seemed very different.

After interviewing participants for the current research, as briefly discussed in the research paper, I attended the eight-week MBCT course in which participants had agreed to take part. Eight out of the nine participants completed the course, which meant I had contact with most participants following the research interviews prior to completing the data analysis. As a result of attending the MBCT group, I felt I gained further insight into participants' experience of psychological distress due to the discussions within the group. Indeed, this was more than would have been gained had I only interviewed the participants and not had any further contact. As such I was able to understand people's experience of psychological distress within the wider context of their life experience, which felt like quite a privileged position to be in. In addition, I also felt like this contributed to ensuring the quality of the research (Yardley, 2000) insofar as I felt I had an increased awareness of the context in which the clients experienced psychological distress.

Furthermore, when I was analysing the data I was aware of my knowledge of psychological distress and therapy from a more general perspective as a trainee clinical psychologist. As such, I was interpreting the data within both the clients' context and my own professional context. Indeed what I did notice was that my knowledge, from a professional perspective, was notably different from that gathered from participants. While I

believe psychological therapy has the potential to improve people's well-being I am aware that this comes from experience of working psychologically and a knowledge of the evidence base in other neurodegenerative diseases. For example, MBCT has previously been piloted with people with Parkinson's disease who found it improved their psychological well-being (Fitzpatrick, Simpson & Smith, 2010). However, it felt important to continually reflect on how my understanding and knowledge was coming from a different place to that of the participants. Indeed, they were coming from a position of lived experience which was the focus of the research.

Consequently, in order to ensure my interpretations were reflective of the data, and not these wider understandings, I aimed as much as possible to bracket off this extra information. Bracketing is a methodological device used in phenomenological inquiry which requires the researcher to deliberately put aside their beliefs and knowledge about the phenomenon under investigation (Carpenter, 2007). This is also to continue throughout the research process. In order to achieve this process I kept a reflective diary whereby I could record and make reflections on the insight I gained from the group that could potentially influence my interpretation of the data. This reflexivity helped to identify potential influences that may have later affected the data analysis and subsequently enabled me to reduce them (Ahern, 1999).

Certainly, during the process of data analysis, I continued to reflect on the interpretations I was making, questioning whether the interpretations being made were based on the data collected or whether the wider knowledge I had gained during the MBCT course was influencing this. I was aware that the additional contact I had with participants could have altered the perspective taken during the data analysis period. For example, as the MBCT course progressed participants talked about finding the course helpful, describing how the how the mindfulness home practice was helping them. As such it felt important to record

these comments and what I had taken from them as the data gathered during my interviews regarding participants' expectations of psychological therapy did not reflect this. Below is an extract from my reflective diary demonstrating this:

During today's session, Lyndsey (pseudonym) talked about how she felt better than she had in a long time. She talked about how she was finding the mindfulness exercises particularly helpful when she is really struggling with her mood. She was really positive about the MBCT course and commented on how she found being in a therapy group with others with HD was comforting. She had been to group therapy before but due to the wide range of reasons for people's attendance did not find the group helpful. This suggests to me that therapy groups, specifically for people with HD, would be of benefit to people.

(Reflective Diary)

Therefore, I did not want to interpret participants' expectations of psychological therapy in a positive light that did not exist. While people were starting to describe the positive impact they felt it was having, during the interviews participants' understandings of psychological therapy were that it provided them with hope but that there were no expectations of whether it would be helpful or not. Consequently, to ensure transparency, a principle in ensuring quality in qualitative research (Yardley, 2000), when writing up the results I presented original quotes from participants transcripts in order to make sure my interpretations were indeed representative of the participants' experiences.

In addition, I used supervision to ensure that the interpretations I had made from the data reflected the participants' direct quotes. One of my supervisors also took part in the MBCT course, therefore it was of particular importance to gain supervision from my second supervisor who did not know the participants and had not had any contact with them

throughout the research process. This ensured that there was a perspective that could not have been influenced by anything other than the data from the research interviews.

### **Impact of time on recruitment**

Indeed, the potential influence attending the group had could have been avoided if I had analysed the data prior to the MBCT course commencing. However, with the current research, timing was problematic. The time between obtaining ethical approval and the commencement of the MBCT course was approximately four weeks. During this time I had to recruit participants and complete all nine interviews. Consequently, I did not have time to transcribe and analyse the data prior to the MBCT course commencing.

Furthermore, there was a wider impact of this time limit in that it also restricted the sample of participants I was able to include in the research. I had initially hoped that partners of those engaging in the MBCT course could be interviewed as well to gain insight into their understanding of their partners' psychological distress and expectations of psychological therapy. It has been demonstrated that people engaged in MBCT have reported an increase in perspective taking and empathy which has subsequently allowed them to interact more mindfully in relationships (Bihari & Mullan, 2014). As such, partners of those with HD may be indirectly affected by their partner taking part in MBCT. I was therefore interested in their perception of their partner engaging in psychological therapy and whether they expected any change as a result of this. It would also have been interesting to be able to see whether people with HD and their partners had similar understandings or whether these diverged.

However, due to the time limit I was unable to recruit enough partners to interview them in time. Indeed, only one partner consented to take part in the research within the time frame I had to complete data collection. I had planned to use IPA (Smith & Osborn, 2003) whereby I would analyse the data of people with HD and their partners separately as two

distinct groups. As such, it did not seem ethical to interview one participant with the potential for their data to be excluded from the research. Therefore, I explained to the partner who had consented to take part my reasons for not interviewing them and including them in the research. Consequently, the priority of the research became to interview people with HD to obtain insight into their experience.

In addition, with regards to future research, it may be important to interview partners to obtain their perspective on psychological distress in HD given the impact it has been shown to have on both the physical and psychological well-being of those around the person with HD (e.g., Aubeeluck, Buchanan & Stupple, 2012; Williams et al., 2009).

### **Absence of psychological services for people with HD**

Interestingly, what became apparent during the interviews, although was not specific to the focus of the present research, was the lack of psychological support available to people with HD. Indeed, if any support were available, this was unknown to the participants. I found it frustrating to learn of the lack of specialist psychological input for people with HD, particularly as the importance of a multi-disciplinary approach in the management of HD and its associated difficulties has been argued (Veenhuizen & Tibben, 2009). Furthermore, it seemed that people were experiencing levels of distress that could be supported by a therapeutic approach. Indeed, many participants talked about not having anyone to share their worries with as they did not want to burden their partners and families. As such they were attempting to cope on their own, with their main contact with health care professionals being at the HD clinic when they attended for review.

I wondered whether the absence of psychological input for people with HD may be, in part, reflective of the dominant biological understanding of psychological distress in HD. Indeed, if people's beliefs regarding their illness are influenced by the information they have

around them (Leventhal, Leventhal & Cameron, 2001) then it seems understandable that people would hold a biological understanding of psychological distress. Consequently, the dominant biological perspective taken would also suggest medication would be the assumed treatment option for people with HD. Indeed, participants spoke about assuming medication was the only option given the biological nature of their distress. As such, if psychological distress is to be understood from an alternative perspective, then the way in which it is talked about should be addressed.

On discussing a psychological approach during the research, participants spoke of their hope that it could improve their psychological well-being. Indeed, this seemed to be in relation to regaining some control over their psychological experience related to HD. However, on reflection, while participants may be able to develop a sense of control, considering the uncontrollable nature of HD, acceptance may also be important for people with HD. Consequently, approaches such as acceptance and commitment therapy (ACT) may support people to manage their psychological distress. Indeed ACT aims to increase a person's psychological flexibility focusing on mindfulness, acceptance and behaviour change in line with a person's values (Hayes, Luoma, Bond, Masuda, & Lillis, 2006). Considering the unchangeable and degenerative nature of HD being able to embrace one's experience without trying to change it, instead accept it, seems important.

### **Impact on self of attending the MBCT course**

Additionally, I felt very grateful to have been able to take part in the MBCT course. This was an experience that is not afforded to many trainee clinical psychologists and I feel this benefitted me from a personal, professional and research perspective. Indeed, I felt I was able to engage with this from both the perspective of a participant and an observer. On the one hand I took part in each session, contributed to group discussion and engaged with the

home practice exercises. From another perspective, being able to observe a qualified clinical psychologist with a wealth of experience in MBCT felt invaluable. I feel I have learned a lot about how MBCT is delivered and what it entails, how to manage group dynamics and the benefits of your own investment in a model. Additionally, since completing the group I have continued with the mindfulness practice at home and maintained the ethos of the model. I believe this has enabled me to manage the demands and stress occurring throughout my thesis journey and training more generally. I have used some of the exercises at times of high stress and others as a means of personal care. I believe this is something I will continue to benefit from throughout my qualified career.

### **Conclusions**

This critical appraisal has been used to reflect on some of the important issues that arose during the research. I have reflected on the interview process and how my role as a trainee clinical psychologist may have influenced this as well as the impact hearing people's stories had on me. I further explored some of the issues that arose around the data analysis process, discussing the potential impact of myself attending the group. Furthermore I explored how time acted as a barrier to my original research proposal resulting in only being able to interview people with HD. However, although having to interview participants within a short space of time resulted in partners being unable to participate I believe this enabled a thorough and detailed understanding of the experiences of people with HD. Finally, I reflected on the lack of psychological services for people with HD, concluding with some reflections around the impact attending the group had on me both personally and professionally.

### References

- Ahern, K.J. (1999). Ten tips for reflexive bracketing. *Qualitative Health Research*, 9 (3), 407-411. doi: 10.1177/104973239900900309.
- Aubeeluck, A., Buchanan, H. & Stupple, E. N. (2012). 'All the burden on all the carers': Exploring quality of life with family caregivers of Huntington's disease patients. *Quality of Life Research*, 21, 1425-1435. doi: 10.1007/s11136-011-0062-x.
- Bihari, J. & Mullan, E. (2014). Relating Mindfully: A Qualitative Exploration of Changes in Relationships Through Mindfulness-Based Cognitive Therapy. *Mindfulness*, 5, 46-59. doi: 10.1007/s12671-012-0146-x.
- Carpenter, D.R. (2007). Phenomenology as method. In H.J. Streubert & D.R. Carpenter (Eds.), *Qualitative research in nursing: Advancing the humanistic imperative* (pp. 75-99). Philadelphia, PA: Lippincott.
- Drury, V., Francis, K., & Chapman, Y. (2007). Taming the rescuer: The therapeutic nature of qualitative research interviews. *International Journal of Nursing Practice*, 13 (6), 383-384. doi: 10.1111/j.1440-172X.2007.00654.x.
- Fitzpatrick, L., Simpson, J. & Smith, A. (2010). A qualitative analysis of mindfulness-based cognitive therapy (MBCT) in Parkinson's disease. *Psychology and Psychotherapy: Theory, Research and Practice*, 83 (2), 179-192. doi: 10.1348/147608309X471514.
- Hayes, S. C., Luoma, J. B., Bond, F. W., Masuda, A., & Lillis, J. (2006). Acceptance and commitment therapy: Model, processes and outcomes. *Behaviour research and therapy*, 44 (1), 1-25. doi: 10.1016/j.brat.2005.06.006.

- Hewitt, J. (2007). Ethical components of research-researched relationships in qualitative interviewing. *Qualitative Health Research, 17* (8), 1149-1159. doi: 10.1177/1049732307308305.
- Leventhal, H., Leventhal, E., & Cameron, L. (2001). Representations, procedures, and affect in illness self-regulation: A perceptual-cognitive model. In Baum, A., Revenson, T., Singer, J. (eds). *Handbook of health psychology*. Hillsdale, NJ: Lawrence Erlbaum Associates (p.19–47).
- Smith, J.A., & Osborn, M. (2003). Interpretative Phenomenological Analysis. In J.A. Smith (Ed.), *Qualitative Psychology*. London: Sage.
- Veenhuizen, R. B., & Tibben, A. (2009). Coordinated multidisciplinary care for Huntington's disease. An outpatient department. *Brain research bulletin, 80* (4), 192-195. doi: 10.1016/j.brainresbull.2009.06.017.
- Wilde, V. (1992). Controversial hypotheses on the relationship between researcher and the informant in qualitative research. *Journal of Advanced Nursing, 17*, 234-242. doi: 10.1111/j.1365-2648.1992.tb01879.x.
- Williams, J. K., Skirton, H., Paulsen, J. S., Tripp-Reimer, T., Jarmon, L., Mcgonigal Kenney, M., Birrer, E., Hennig, B. L. & Honeyford, J. (2009). The emotional experiences of family carers in Huntington disease. *Journal of Advanced Nursing, 65*, 789-798. doi: 10.1111/j.1365-2648.2008.04946.x.
- Yardley, L. (2000). Dilemmas in qualitative health research. *Psychology and Health, 15* (2), 215-228. doi: 10.1080/08870440008400302.

## **Section 4: Ethics**

Rachael Theed

Trainee Clinical Psychologist

Doctorate in Clinical Psychology

Division of Health Research, Lancaster University

Word Count: 4752

All correspondence should be sent to:

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Email: [r.theed@lancaster.ac.uk](mailto:r.theed@lancaster.ac.uk)

## Ethics Application Form

NHS REC Form

Reference:  
15/YH/0377

IRAS Version 5.0.0

Welcome to the Integrated Research Application System

IRAS Project Filter

The integrated dataset required for your project will be created from the answers you give to the following questions. The system will generate only those questions and sections which (a) apply to your study type and (b) are required by the bodies reviewing your study. Please ensure you answer all the questions before proceeding with your applications.

Please complete the questions in order. If you change the response to a question, please select 'Save' and review all the questions as your change may have affected subsequent questions.

Please enter a short title for this project (maximum 70 characters)  
Expectations of psychological therapy in Huntington's disease

1. Is your project research?

Yes  No

2. Select one category from the list below:

- Clinical trial of an investigational medicinal product
- Clinical investigation or other study of a medical device
- Combined trial of an investigational medicinal product and an investigational medical device
- Other clinical trial to study a novel intervention or randomised clinical trial to compare interventions in clinical practice
- Basic science study involving procedures with human participants
- Study administering questionnaires/interviews for quantitative analysis, or using mixed quantitative/qualitative methodology
- Study involving qualitative methods only
- Study limited to working with human tissue samples (or other human biological samples) and data (specific project only)
- Study limited to working with data (specific project only)
- Research tissue bank
- Research database

If your work does not fit any of these categories, select the option below:

Other study

2a. Please answer the following question(s):

- a) Does the study involve the use of any ionising radiation?  Yes  No
- b) Will you be taking new human tissue samples (or other human biological samples)?  Yes  No
- c) Will you be using existing human tissue samples (or other human biological samples)?  Yes  No

3. In which countries of the UK will the research sites be located? (Tick all that apply)

- England
- Scotland
- Wales
- Northern Ireland

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- England  
 Scotland  
 Wales  
 Northern Ireland  
 This study does not involve the NHS

4. Which review bodies are you applying to?

- HRA Approval  
 NHS/HSC Research and Development offices  
 Social Care Research Ethics Committee  
 Research Ethics Committee  
 Confidentiality Advisory Group (CAG)  
 National Offender Management Service (NOMS) (Prisons & Probation)

*For NHS/HSC R&D offices, the CI must create Site-Specific Information Forms for each site, in addition to the study-wide forms, and transfer them to the PIs or local collaborators.*

5. Will any research sites in this study be NHS organisations?

- Yes  No

6. Do you plan to include any participants who are children?

- Yes  No

7. Do you plan at any stage of the project to undertake intrusive research involving adults lacking capacity to consent for themselves?

- Yes  No

*Answer Yes if you plan to recruit living participants aged 16 or over who lack capacity, or to retain them in the study following loss of capacity. Intrusive research means any research with the living requiring consent in law. This includes use of identifiable tissue samples or personal information, except where application is being made to the Confidentiality Advisory Group to set aside the common law duty of confidentiality in England and Wales. Please consult the guidance notes for further information on the legal frameworks for research involving adults lacking capacity in the UK.*

8. Do you plan to include any participants who are prisoners or young offenders in the custody of HM Prison Service or who are offenders supervised by the probation service in England or Wales?

- Yes  No

9. Is the study or any part of it being undertaken as an educational project?

- Yes  No

Please describe briefly the involvement of the student(s):  
The student will be the principal investigator.

9a. Is the project being undertaken in part fulfilment of a PhD or other doctorate?

- Yes  No

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10. Will this research be financially supported by the United States Department of Health and Human Services or any of its divisions, agencies or programs?

Yes  No

11. Will identifiable patient data be accessed outside the care team without prior consent at any stage of the project (including identification of potential participants)?

Yes  No

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NHS REC Form

Reference:  
15/YH/0377

IRAS Version 5.0.0

Integrated Research Application System  
Application Form for Research involving qualitative methods only



Application to NHS/HSC Research Ethics Committee

The Chief Investigator should complete this form. Guidance on the questions is available wherever you see this symbol displayed. We recommend reading the guidance first. The complete guidance and a glossary are available by selecting [Help](#).

Please define any terms or acronyms that might not be familiar to lay reviewers of the application.

Short title and version number: (maximum 70 characters - this will be inserted as header on all forms)  
Expectations of psychological therapy in Huntington's disease

Please complete these details after you have booked the REC application for review.

REC Name:  
[Redacted]

REC Reference Number: [Redacted]      Submission date: 27/07/2015

**PART A: Core study information**

**1. ADMINISTRATIVE DETAILS**

A1. Full title of the research:  
Understandings of psychological difficulties in Huntington's disease and expectations of psychological therapy

A2-1. Educational projects

Name and contact details of student(s):

Student 1			
	Title	Forename/Initials	Surname
		Miss Rachael	Theed
Address	Furness College Lancaster University Lancaster		
Post Code	LA1 4YT		
E-mail	r.theed@lancaster.ac.uk		
Telephone	07952904313		
Fax			
Give details of the educational course or degree for which this research is being undertaken:			

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Name and level of course/ degree :  
Doctorate in Clinical Psychology

Name of educational establishment:  
Lancaster University

Name and contact details of academic supervisor(s):

Academic supervisor 1

	Title	Forename/Initials	Surname
	Dr	Jane	Simpson
Address	Doctorate in Clinical Psychology Furness College, Lancaster University Lancaster		
Post Code	LA1 4YT		
E-mail	j.simpson2@lancaster.ac.uk		
Telephone	015 2459 2858		
Fax			

Academic supervisor 2

	Title	Forename/Initials	Surname
	Dr	Fiona	Eccles
Address	Doctorate in Clinical Psychology Furness College, Lancaster University Lancaster		
Post Code	LA1 4YT		
E-mail	feccles@lancaster.ac.uk		
Telephone	015 2459 2807		
Fax			

Please state which academic supervisor(s) has responsibility for which student(s):  
Please click "Save now" before completing this table. This will ensure that all of the student and academic supervisor details are shown correctly.

Student(s)	Academic supervisor(s)
Student 1 Miss Rachael Theed	<input checked="" type="checkbox"/> Dr Jane Simpson <input checked="" type="checkbox"/> Dr Fiona Eccles

A copy of a current CV for the student and the academic supervisor (maximum 2 pages of A4) must be submitted with the application.

A2-2. Who will act as Chief Investigator for this study?

Student  
 Academic supervisor  
 Other

A3-1. Chief Investigator:

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	Title Forename/Initials Surname
	Miss Rachael Theed
Post	Trainee Clinical Psychologist
Qualifications	BSc(Hons) MSc
Employer	Lancashire Care Foundation Trust
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Post Code	LA1 4YT
Work E-mail	r.theed@lancaster.ac.uk
* Personal E-mail	
Work Telephone	07508 406193
* Personal Telephone/Mobile	
Fax	

*\* This information is optional. It will not be placed in the public domain or disclosed to any other third party without prior consent.  
A copy of a current CV (maximum 2 pages of A4) for the Chief Investigator must be submitted with the application.*

Who is the contact on behalf of the sponsor for all correspondence relating to applications for this project?  
This contact will receive copies of all correspondence from REC and HRA/R&D reviewers that is sent to the CI.

	Title Forename/Initials Surname
	Ms Debbie Knight
Address	Research Support Officer B58 Bowland Main Lancaster University
Post Code	LA1 4YT
E-mail	ethics@lancaster.ac.uk
Telephone	01524592605
Fax	

A5-1. Research reference numbers. Please give any relevant references for your study:

Applicant's/organisation's own reference number, e.g. R & D (if available):

Sponsor's/protocol number:

Protocol Version: Version 1

Protocol Date: 22.06.2015

Funder's reference number:

Project website:

Additional reference number(s):

Ref.Number	Description	Reference Number

*Registration of research studies is encouraged wherever possible. You may be able to register your study through your NHS organisation or a register run by a medical research charity, or publish your protocol through an open access publisher. If you have registered your study please give details in the "Additional reference number(s)" section.*

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A5-2. Is this application linked to a previous study or another current application?

 Yes  No

Please give brief details and reference numbers.

This research is linked to "A pilot evaluation of mindfulness-based cognitive therapy for people with Huntington's disease".

REC reference: [REDACTED]  
 IRAS ID: [REDACTED]  
 ClinicalTrials.gov ID: [REDACTED]  
 Chief Investigator: Dr Jane Simpson

## 2. OVERVIEW OF THE RESEARCH

To provide all the information required by review bodies and research information systems, we ask a number of specific questions. This section invites you to give an overview using language comprehensible to lay reviewers and members of the public. Please read the guidance notes for advice on this section.

**A6-1. Summary of the study.** Please provide a brief summary of the research (maximum 300 words) using language easily understood by lay reviewers and members of the public. Where the research is reviewed by a REC within the UK Health Departments' Research Ethics Service, this summary will be published on the Health Research Authority (HRA) website following the ethical review. Please refer to the question specific guidance for this question.

A pilot trial of mindfulness-based cognitive therapy (MBCT) with people with Huntington's disease (HD) is due to take place later this year (clinicaltrials.gov ICT [REDACTED]). The current study aims to explore the hopes and expectations of some of those who are due to take part in this intervention and their partners. A small group of people with HD and their partners will be interviewed about their understanding of psychological distress and their beliefs and expectations of psychological therapy and MBCT specifically. The data will be analysed using interpretative phenomenological analysis.

**A6-2. Summary of main issues.** Please summarise the main ethical, legal, or management issues arising from your study and say how you have addressed them.

Not all studies raise significant issues. Some studies may have straightforward ethical or other issues that can be identified. Others may present significant issues requiring further consideration by a REC, R&D office or other review body (as appropriate to the issue). Studies that present a minimal risk to participants may raise complex organisational or legal issues. You should try to consider all the types of issues that the different reviewers may need to consider.

### Ethical Issues

1. Confidentiality: All participant information will remain confidential. The researcher will not have access to potential participants' personal information until the participants themselves express an interest in participating in the study and provide their own contact details. The researcher and her supervisors will be the only people to have access to the audio recordings and transcripts. This will be outlined on the participant information sheet and consent form.

2. Risk of harm to participants: While it is not expected that the research will cause participants any distress, if this occurs during interviews the participant will be given a break and asked if they would like to continue. Should any of the participants disclose any risk of harm to themselves or others then this will be disclosed to their treating clinician or other appropriate person or authorities. Where possible participants will be informed if this is necessary.

3. Risk of harm to the researcher: As the interviews will be conducted on a one to one basis, when lone working the researcher will follow the Lancashire Care Foundation Trust lone working policy. A designated person (a fellow trainee) will be provided with the researcher's contact details and information about the appointment time and duration. They will further be provided with contact details of the interview location and interviewee (in a sealed envelope, only to be opened in an emergency). When the interview is finished the researcher will contact the designated person to inform them that the interview is complete. If they do not receive this contact then attempts will be made to contact the researcher. If they are unable to make contact then they can open the sealed envelope and the appropriate authorities will be informed.

4. Data Storage: All electronic data will be stored on the University server via the VPN and password protected. All

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paper data will be stored in a locked draw at the home of the researcher during the course of the research.

A6-3. Proportionate review of REC application. *The initial project filter has identified that your study may be suitable for proportionate review by a REC sub-committee. Please consult the current guidance notes from NRES and indicate whether you wish to apply through the proportionate review service or, taking into account your answers to A6-2, you consider there are ethical issues that require consideration at a full REC meeting.*

Yes - proportionate review  No - review by full REC meeting

*Further comments (optional):*

*Note: This question only applies to the REC application.*

### 3. PURPOSE AND DESIGN OF THE RESEARCH

A7. Select the appropriate methodology description for this research. *Please tick all that apply.*

- Case series/ case note review
- Case control
- Cohort observation
- Controlled trial without randomisation
- Cross-sectional study
- Database analysis
- Epidemiology
- Feasibility/pilot study
- Laboratory study
- Metanalysis
- Qualitative research
- Questionnaire, interview or observation study
- Randomised controlled trial
- Other (please specify)

A10. What is the principal research question/objective? *Please put this in language comprehensible to a lay person.*

The main objective is to gain insight into both people with Huntington's disease and their partners' understanding of psychological distress and their views/expectations of psychological therapy, in particular mindfulness-based cognitive therapy.

A11. What are the secondary research questions/objectives if applicable? *Please put this in language comprehensible to a lay person.*

What impact has psychological therapy had/do people expect it to have on both the person with Huntington's disease and their partners emotional and psychological wellbeing?

A12. What is the scientific justification for the research? *Please put this in language comprehensible to a lay person.*

Huntington's disease (HD) is a neurodegenerative condition causing problems with cognitive functioning, coordination, movement and some emotional difficulties. It is suggested that around five to ten per 100,000 people are affected (Kay, Fisher & Hayden, 2014). People are generally diagnosed between the ages of 35-55 with a life expectancy of around 15-20 years (Keenan, Simpson, Miedzybrodzka, Alexander & Semper, 2013) and have often seen their parents affected by the disease (Kremer, 2002).

People with HD often experience emotional difficulties including depression, anxiety, apathy and irritability (Kirkwood, Su, Conneally & Foroud, 2001) which have the potential to impact on an individual's quality of life. For example, rates

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of depression have been shown to be high in individuals with HD with the causes for this being potentially twofold (Paulsen et al., 2005). Certainly one potential cause is having to adjust to living with the illness as well as coping with the subsequent difficulties that arise as a result of HD. Additionally, there is a potential biological component whereby neural mechanisms in the brain are affected by HD subsequently affecting mood (Paulsen et al., 2005).

Importantly, there are significant physical and emotional consequences of HD not only for the individual with the disease but also for the people with whom they live (e.g., Aubeeluck, Buchanan & Stupple, 2012; Williams et al., 2009). Family members often take on caregiving for the individual with HD and subsequently may experience a reduced quality of life, including lowered mood (Aubeeluck & Buchanan, 2007; McCabe, Firth & O'Connor, 2009; Read et al., 2010), potentially as a result of witnessing a loved one become increasingly unwell over time and grieving for a lost relationship (Pickett, Altmajer & Paulsen, 2007). Indeed, depression in people with HD has been identified as being a strong predictor of depression in those caring for someone with HD (Pickett et al., 2007).

Although for people living with HD medication is often the main approach in supporting people with mood difficulties,

preferred option for people with HD and there could be alternative options. For example, in another neurodegenerative disease, Parkinson's disease (PD), while medication may be used to support people with the psychological difficulties they experience, there is increasing evidence to support the use of psychological interventions in this population (Charidimou, Seamons, Selai & Schrag, 2011; Dobkins et al., 2011), including mindfulness-based cognitive therapy (MBCT; Fitzpatrick, Simpson & Smith, 2010). Due to the level of distress both people with HD and their partners experience, it is important to understand what people think about the support available and treatment they receive.

MBCT is an eight week group therapy developed by Segal, Williams and Teasdale (2002) which teaches mindfulness skills through a range of practices with the aim of preventing the reoccurrence of depression (Gu, Strauss, Bond & Cavanagh, 2015). Previously MBCT has been piloted with people with Parkinson's disease who found it improved their psychological well-being (Fitzpatrick, Simpson & Smith, 2010). MBCT is therefore going to be piloted for individuals with HD with the aim of alleviating psychological distress directly in people with HD and indirectly in their partners. Although the partners will not be directly participating in the MBCT groups, higher levels of mindfulness have been shown to be associated with higher levels of satisfaction in partner relationships (Barnes, Brown, Krusemark, Campbell & Rogge, 2007). Furthermore, people engaged in MBCT have reported increased empathy and perspective taking, allowing them to respond more mindfully in relationships (Bihari & Mullan, 2014).

Consequently, this study aims to investigate both individuals with HD and their partners' understanding of psychological difficulties in HD and their views of psychological therapy. Participants will be recruited from those who are due to take part in the trial of MBCT and therefore the study will focus in particular on mindfulness and people's knowledge of this, as well as their hopes and expectations for MBCT. This study will adopt a qualitative methodology to obtain detailed accounts of people with HD and their partners understanding of psychological difficulties and expectations of psychological therapy. In order to address this, semi-structured interviews will be conducted and analysed by interpretative phenomenological analysis (IPA; Smith, 2009).

Aubeeluck, A., Buchanan, H. & Stupple, E. N. (2012). 'All the burden on all the carers': exploring quality of life with family caregivers of Huntington's disease patients. *Quality of Life Research*, 21, 1425-1435.

Aubeeluck, A. & Buchanan, H. (2007). The Huntington's Disease quality of life battery for carers: reliability and validity. *Clinical Genetics*, 71, 434-445.

Barnes, S., Brown, K. W., Krusemark, E., Campbell, W. K. & Rogge, R. D. (2007). The role of mindfulness in romantic relationship satisfaction and responses to relationship stress. *Journal of Marital and Family Therapy*, 33 (4), 482-500.

Bihari, J. & Mullan, E. (2014). *Relating Mindfully: A Qualitative Exploration of Changes in Relationships Through Mindfulness-Based Cognitive Therapy*. *Mindfulness*, 5, 46-59.

Charidimou, A., Seamons, J., Selai, C., & Schrag, A. (2011). The role of Cognitive-Behavioural Therapy for patients with depression in Parkinson's disease. *Parkinson's disease*, 2011.

Dobkin, R.D., Menza, M., Allen, L.A., Gara, M.A., Mark, M.H., Tiu, J., ... Friedman, J. (2011). Cognitive behavioural therapy for depression in Parkinson's disease: A randomised, controlled trial. *American Journal of Psychiatry*, 168 (10), 1066-1074.

Fitzpatrick, L., Simpson, J. & Smith, A. (2010). A qualitative analysis of mindfulness-based cognitive therapy (MBCT) in Parkinson's disease. *Psychology and Psychotherapy: Theory, Research and Practice*, 83 (2), 179-192.

Gu, J., Strauss, C., Bond, R., & Cavanagh, K. (2015). How do mindfulness-based cognitive therapy and mindfulness-based stress reduction improve mental health and well-being? A systematic review and meta-analysis of mediation studies. *Clinical Psychology Review*, 37, 1-12.

Kay, C., Fisher, E. & Hayden, M. R. 2014. Epidemiology. In: Bates, G., Tabrizi, S. J. & Jones, L. (eds.) *Huntington's disease*. 4th ed. Oxford: Oxford University Press.

Keenan, K.F., Simpson, S.A., Medzybrodzka, Z., Alexander, D.A., & Semper, J. (2013). How do partners find out about the risk of Huntington's disease in couple relationships. *Journal of Genetic Counselling*, 22, 336-344.

Kirkwood, S.C., Su, J.L., Conneally, M., & Foroud, T. (2001). Progression of symptoms in the early and middle stages of Huntington's disease. *Archives of Neurology*, 58 (2), 273-278.

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Kremer, B. (2002). Clinical neurology in Huntington's disease. In: Bates, G., Harper, P. & Jones, L. (eds.) Huntington's disease. 3rd ed. Oxford: Oxford University Press.

Mccabe, M. P., Firth, L. & O'connor, E. (2009). A Comparison of Mood and Quality of Life Among People with Progressive Neurological Illnesses and Their Caregivers. *Journal of Clinical Psychology in Medical Settings*, 16, 355-362.

Paulsen, J.S., Nehl, C., Hoth, K.F., Kanz, J.E., Benjamin, M., Conybeare, R., ... & Turner, B. (2005). Depression and stages of Huntington's disease. *Journal of Neuropsychiatry and Clinical Neurosciences*, 17 (4), 496-502.

Pickett, T., Altmaier, E., & Paulsen, J.S. (2007). Caregiver burden in Huntington's disease. *Rehabilitation Psychology*, 52 (3), 311-318.

Read, J. E., A. T. P., Langbehn, D. R., Owen, G., Dürr, A., Leavitt, B. R., Roos, R. a. C., Stout, J. C., Tabrizi, S. J., Craufurd, D. & Investigators, T.-H. (2010). Quality of life in Huntington's disease: a comparative study investigating the impact on spouses of those with premanifest and early disease. *Journal of Neurology, Neurosurgery & Psychiatry*, 81, A23-A24.

Segal, Z.V., Williams, J.M., & Teasdale, J. (2002). *Mindfulness-based cognitive therapy for depression: A new approach to preventing relapse*. London, UK: Guildford Press.

Smith, J. A. (2009). *Interpretative phenomenological analysis: theory, method and research*. London, Sage.

Williams, J. K., Skirton, H., Paulsen, J. S., Tripp-Reimer, T., Jarmon, L., Magonigal Kenney, M., Birner, E., Hennig, B. L. & Honeyford, J. (2009). The emotional experiences of family carers in Huntington disease. *Journal of Advanced Nursing*, 65, 789-798.

A13. Please summarise your design and methodology. *It should be clear exactly what will happen to the research. Please complete this section in language comprehensible to the lay person. Do not simply reproduce or refer to the protocol. Further guidance is available in the guidance notes.*

#### Design

This study will collect qualitative data from both people with Huntington's disease (HD) and partners of people with HD.

Semi-structured interviews will be conducted with people with HD and partners of people with HD prior to them engaging in a mindfulness-based cognitive therapy trial. Interviews will be analysed by interpretative phenomenological analysis (IPA; Smith, 2009) with the data being analysed separately for people with HD and their partners. IPA is widely used in psychological research and explores how a homogenous group of people understand and make sense of their experiences (Smith, Flowers & Larkin, 2009). However, if there are insufficient participants (either people with HD or partners of people with HD) then the data will be analysed using thematic analysis (Braun & Clarke, 2006).

#### Method

People with HD will be recruited from the MBCT trial. Potential participants will be informed about the research when they have opted into the MBCT trial. Those suitable for inclusion in the study will be provided with an information pack including a participant information sheet (appendix 3) and consent to contact form (appendix 4) either via post, email or in person by the recruiters for the main MBCT trial (Dr Fiona Eccles & Dr Jane Simpson) with a cover letter (appendix 5). As partners will be recruited through the person with HD engaging in the MBCT programme, if they are interested, an information pack for the partner will be sent to the person with HD via email or post. They will then be asked to pass this on to their partner. The information pack will contain an information sheet and consent to contact form. They will then be asked to contact the researcher (via the consent to contact form, email or telephone) for an initial discussion regarding the research to consider whether they would like to take part.

Due to the study only requiring a small number of participants there is the potential for people to opt into the study once the target has been reached. In this instance, potential participants will be contacted by the researcher informing them that the target sample has been met.

If people with HD and their partners decide to take part in the research then a mutually convenient interview time will be arranged. Participants will be interviewed individually either at home or at a community location convenient for the participant. People's partners do not have to take part in order for the person with HD to take part and vice versa. However, if the person with HD decides not to participate then partner will still be recruited in the same way via the person with HD. Before commencing the interview the researcher will check each participant has read the participant information sheet and go through the consent form (appendix 6 & 7), answering any questions participants may have. Participants will be interviewed prior to the person with HD engaging in the MBCT programme. Interviews are anticipated to last approximately 60 minutes per person. At the end of the interview participants will be debriefed using the debrief sheet (appendix 8) containing sources of support should they require this.

Telephone interviews may also be conducted if participants are not able to meet. If this is the case, a consent form will be posted to the participant and returned prior to the interview. Participants will be debriefed over the phone as well as having the debrief sheet posted or emailed to them following the interview.

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A14-1. In which aspects of the research process have you actively involved, or will you involve, patients, service users, and/or their carers, or members of the public?

- Design of the research
- Management of the research
- Undertaking the research
- Analysis of results
- Dissemination of findings
- None of the above

*Give details of involvement, or if none please justify the absence of involvement.*  
Advice was sought from the Huntington's disease association in the design of the materials for the main MBCT trial and the same advice was followed in the design of these materials.

4. RISKS AND ETHICAL ISSUES

RESEARCH PARTICIPANTS

A17-1. Please list the principal inclusion criteria (list the most important, max 5000 characters).

To participate in the present study participants must meet the following criteria:  
 - People with HD will be signed up to participate in the MBCT trial.  
 - Partners of those signed up to participate in the MBCT trial.  
 - Participants must be aged 18 or over.

A17-2. Please list the principal exclusion criteria (list the most important, max 5000 characters).

People with HD and partners of someone with HD not enrolled on to the MBCT trial will not be eligible for inclusion.

RESEARCH PROCEDURES, RISKS AND BENEFITS

A18. Give details of all non-clinical intervention(s) or procedure(s) that will be received by participants as part of the research protocol. These include seeking consent, interviews, non-clinical observations and use of questionnaires.

Please complete the columns for each intervention/procedure as follows:

1. Total number of interventions/procedures to be received by each participant as part of the research protocol.
2. If this intervention/procedure would be routinely given to participants as part of their care outside the research, how many of the total would be routine?
3. Average time taken per intervention/procedure (minutes, hours or days)
4. Details of who will conduct the intervention/procedure, and where it will take place.

Intervention or procedure	1	2	3	4
Introduction of research study (person with HD and partner)	1	10		Face to face conversation with Fiona Eccles or through the post.
Receive an email or post information pack and consent to contact form	1	5		Rachael Theed
Initial conversation (people with HD and partners)	1	15		Telephone conversation conducted by Rachael Theed
Provide written consent	1	15		Taking of informed consent by Rachael Theed at a location convenient for the participant
Be interviewed prior to engaging in the	1	60		Participant will be interviewed by Rachael Theed either at their

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home or a community location convenient to them or via  
telephone

A21. How long do you expect each participant to be in the study in total?

Participants will opt in to the study approximately between July-September and will have been interviewed by the end of October 2015. Therefore each participant will be in the study for approximately three months.

A22. What are the potential risks and burdens for research participants and how will you minimise them?

*For all studies, describe any potential adverse effects, pain, discomfort, distress, intrusion, inconvenience or changes to lifestyle. Only describe risks or burdens that could occur as a result of participation in the research. Say what steps would be taken to minimise risks and burdens as far as possible.*

During the interviews there is the potential for participants to become distressed due to the sensitive nature of the research. Should this occur, participants will be given the opportunity to take a break and asked whether they would like to continue or not. They will be debriefed at the end of the interview and provided with a list of contacts for support should they feel this necessary.

A23. Will interviews/questionnaires or group discussions include topics that might be sensitive, embarrassing or upsetting, or is it possible that criminal or other disclosures requiring action could occur during the study?

Yes  No

*If Yes, please give details of procedures in place to deal with these issues:*

If a participant become distressed during the interview then they will be given a break and the researcher will check whether they wish to continue or not. Participants will be debriefed after the interview and provided with a contact list of support services should they feel this necessary. If there is concern that a participant poses a risk of harm, either to themselves or others, then the researcher will inform [REDACTED] (participant with HD's treating clinician) or other appropriate authorities.

A24. What is the potential for benefit to research participants?

There is no direct benefit to participants. However, the insight gained into people's experiences of psychological therapy and expectations/hopes for therapy will contribute to the evidence base for the need for psychological therapies to be widely available for people with HD.

A26. What are the potential risks for the researchers themselves? (if any)

When lone working the researcher will follow the Lancashire Care Foundation Trust lone working policy. A designated person (a fellow trainee) will be provided with the researcher's contact details and information about the appointment time and duration. They will further be provided with contact details of the interview location and interviewee (in a sealed envelope, only to be opened in an emergency). When the interview is finished the researcher will contact the designated person to inform them that the interview is complete. If they do not receive this contact then attempts will be made to contact the researcher. If they are unable to make contact then they can open the sealed envelope and the appropriate authorities will be informed.

#### RECRUITMENT AND INFORMED CONSENT

*In this section we ask you to describe the recruitment procedures for the study. Please give separate details for different study groups where appropriate.*

A27-1. How will potential participants, records or samples be identified? Who will carry this out and what resources will be used? For example, identification may involve a disease register, computerised search of GP records, or review of medical records. Indicate whether this will be done by the direct healthcare team or by researchers acting under arrangements with the responsible care organisation(s).

Participants with Huntington's disease will have been identified and recruited into the MBCT trial by the clinical team at [REDACTED] then initially approached by either Dr Fiona Eccles or Dr [REDACTED]

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will then be sent an information pack and consent to contact form should they wish to know more about the research or opt in.

A27-2. Will the identification of potential participants involve reviewing or screening the identifiable personal information of patients, service users or any other person?

Yes  No

Please give details below:

Participants will already have been screened to engage in the MBCT trial.

A28. Will any participants be recruited by publicity through posters, leaflets, adverts or websites?

Yes  No

A29. How and by whom will potential participants first be approached?

Potential participants will initially be approached by either Dr Fiona Eccles or Dr Jane Simpson when discussing the MBCT trial. Fiona Eccles will provide them with an information pack and consent to contact form if they wish to discuss the study further or opt in.

A30-1. Will you obtain informed consent from or on behalf of research participants?

Yes  No

*If you will be obtaining consent from adult participants, please give details of who will take consent and how it will be done, with details of any steps to provide information (a written information sheet, video, or interactive material). Arrangements for adults unable to consent for themselves should be described separately in Part B Section 6, and for children in Part B Section 7.*

*If you plan to seek informed consent from vulnerable groups, say how you will ensure that consent is voluntary and fully informed.*

Informed written consent will be obtained by Rachael Theed. Participants will have been provided with a participant information sheet explaining the purpose of the study and what the study will involve. Written consent will be obtained face to face where possible prior to conducting the interviews. In the case of telephone interviews, participants will have been posted or emailed a consent form which will need to be returned prior to conducting the interview.

*If you are not obtaining consent, please explain why not.*

Please enclose a copy of the information sheet(s) and consent form(s).

A30-2. Will you record informed consent (or advice from consultees) in writing?

Yes  No

A31. How long will you allow potential participants to decide whether or not to take part?

Participants will have a couple of weeks to decide whether or not they would like to take part. They will be given the information in August 2015 and will have until the beginning of September 2015 to decide due to the interviews needing to be conducted prior to the MBCT course which is anticipated to commence in October 2015.

A33-1. What arrangements have been made for persons who might not adequately understand verbal explanations or written information given in English, or who have special communication needs? (e.g. translation, use of interpreters)

Due to participants needing to understand verbal instructions and explanations given in English during the MBCT trial,

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potential participants in this study will understand English. Those who do not will have been excluded prior to being approached to take part in this research.

A35. What steps would you take if a participant, who has given informed consent, loses capacity to consent during the study? *Tick one option only.*

- The participant and all identifiable data or tissue collected would be withdrawn from the study. Data or tissue which is not identifiable to the research team may be retained.
- The participant would be withdrawn from the study. Identifiable data or tissue already collected with consent would be retained and used in the study. No further data or tissue would be collected or any other research procedures carried out on or in relation to the participant.
- The participant would continue to be included in the study.
- Not applicable – informed consent will not be sought from any participants in this research.
- Not applicable – it is not practicable for the research team to monitor capacity and continued capacity will be assumed.

*Further details:*

*If you plan to retain and make further use of identifiable data/tissue following loss of capacity, you should inform participants about this when seeking their consent initially.*

#### CONFIDENTIALITY

In this section, personal data means any data relating to a participant who could potentially be identified. It includes pseudonymised data capable of being linked to a participant through a unique code number.

#### Storage and use of personal data during the study

A36. Will you be undertaking any of the following activities at any stage (including in the identification of potential participants)? *(Tick as appropriate)*

- Access to medical records by those outside the direct healthcare team
- Electronic transfer by magnetic or optical media, email or computer networks
- Sharing of personal data with other organisations
- Export of personal data outside the EEA
- Use of personal addresses, postcodes, faxes, emails or telephone numbers
- Publication of direct quotations from respondents
- Publication of data that might allow identification of individuals
- Use of audio/visual recording devices
- Storage of personal data on any of the following:
- Manual files including X-rays
  - NHS computers
  - Home or other personal computers
  - University computers
  - Private company computers
  - Laptop computers

*Further details:*

A38. How will you ensure the confidentiality of personal data? *Please provide a general statement of the policy and*

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*procedures for ensuring confidentiality, e.g. anonymisation or pseudonymisation of data.*

All personal data will be confidential. Each participant will be given a pseudonym to ensure all quotes are not identifiable. Furthermore, all interview data will be anonymised as far as possible.

A40. Who will have access to participants' personal data during the study? Where access is by individuals outside the direct care team, please justify and say whether consent will be sought.

The principal researcher and her supervisors will have access to participants' personal data. This will have been provided by each participant at the beginning of the research.

#### Storage and use of data after the end of the study

A43. How long will personal data be stored or accessed after the study has ended?

- Less than 3 months  
 3 – 6 months  
 6 – 12 months  
 12 months – 3 years  
 Over 3 years

*If longer than 12 months, please justify:*

Consent forms will be scanned and stored on the university server for 10 years. Paper copies will be destroyed at this point. All other personal data will be destroyed once the research has been submitted. However, if participants request a copy of the results then their contact details will be kept until they have been sent out.

#### INCENTIVES AND PAYMENTS

A46. Will research participants receive any payments, reimbursement of expenses or any other benefits or incentives for taking part in this research?

- Yes  No

A47. Will individual researchers receive any personal payment over and above normal salary, or any other benefits or incentives, for taking part in this research?

- Yes  No

A48. Does the Chief Investigator or any other investigator/collaborator have any direct personal involvement (e.g. financial, share holding, personal relationship etc.) in the organisations sponsoring or funding the research that may give rise to a possible conflict of interest?

- Yes  No

#### NOTIFICATION OF OTHER PROFESSIONALS

A49-1. Will you inform the participants' General Practitioners (and/or any other health or care professional responsible for their care) that they are taking part in the study?

- Yes  No

*If Yes, please enclose a copy of the information sheet/letter for the GP/health professional with a version number and date.*

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## PUBLICATION AND DISSEMINATION

Will the research be registered on a public database?

 Yes  No

Please give details, or justify if not registering the research.  
The main MBCT trial is registered.

ClinicalTrials.gov ID [REDACTED]

Registration of research studies is encouraged wherever possible.  
You may be able to register your study through your NHS organisation or a register run by a medical research charity,  
or publish your protocol through an open access publisher. If you are aware of a suitable register or other method of  
publication, please give details. If not, you may indicate that no suitable register exists. Please ensure that you have  
entered registry reference number(s) in question A5-1.

A51. How do you intend to report and disseminate the results of the study? Tick as appropriate:

- Peer reviewed scientific journals  
 Internal report  
 Conference presentation  
 Publication on website  
 Other publication  
 Submission to regulatory authorities  
 Access to raw data and right to publish freely by all investigators in study or by Independent Steering Committee  
on behalf of all investigators  
 No plans to report or disseminate the results  
 Other (please specify)

A53. Will you inform participants of the results?

 Yes  No

Please give details of how you will inform participants or justify if not doing so.  
Participants will be sent a copy of the results if they request them.

## 5. Scientific and Statistical Review

A54. How has the scientific quality of the research been assessed? Tick as appropriate:

- Independent external review  
 Review within a company  
 Review within a multi-centre research group  
 Review within the Chief Investigator's institution or host organisation  
 Review within the research team  
 Review by educational supervisor  
 Other

Justify and describe the review process and outcome. If the review has been undertaken but not seen by the  
researcher, give details of the body which has undertaken the review:

The research has been reviewed by the Lancaster University research tutors.

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*For all studies except non-doctoral student research, please enclose a copy of any available scientific critique reports, together with any related correspondence.*

*For non-doctoral student research, please enclose a copy of the assessment from your educational supervisor/institution.*

A59. What is the sample size for the research? How many participants/samples/data records do you plan to study in total? If there is more than one group, please give further details below.

Total UK sample size: 24  
Total international sample size (including UK):  
Total in European Economic Area:

Further details:

A60. How was the sample size decided upon? If a formal sample size calculation was used, indicate how this was done, giving sufficient information to justify and reproduce the calculation.

This is a sufficient number of participants to analyse the data using interpretative phenomenological analysis (IPA; Smith, 2009).

A62. Please describe the methods of analysis (statistical or other appropriate methods, e.g. for qualitative research) by which the data will be evaluated to meet the study objectives.

Following each interview, the data will be transcribed. Interpretative phenomenological analysis will be used to analyse the data, following the stages outlined by Smith (2009). IPA is widely used in psychological research and aims to explore how homogenous groups of people understand and make sense of their personal and social world (Smith, Flowers & Larkin, 2009). Interviews will be transcribed and analysed individually, in turn. Transcripts will be read and re-read with initial notations relating to the data being made in the margin which will be used to identify potential themes across transcripts. Data will then be organised into superordinate themes including sub-themes where appropriate.

Smith, J. A. (2009). Interpretative phenomenological analysis: theory, method and research. London, Sage.  
Smith, J. A., Flowers, P. & Larkin, M. (2009). Interpretative phenomenological analysis: theory method and research: understanding method and application. London, Sage.

## 6. MANAGEMENT OF THE RESEARCH

A63. Other key investigators/collaborators. Please include all grant co-applicants, protocol co-authors and other key members of the Chief Investigator's team, including non-doctoral student researchers.

	Title Forename/Initials Surname
	Dr Jane Simpson
Post	Research Director, DclinPsy and Director of Education for DHR
Qualifications	BA(Hons), BSc, PhD, DClInPsy, CPsychol
Employer	Lancaster University
Work Address	Doctorate in Clinical Psychology Furness College, Lancaster University Lancaster
Post Code	LA1 4YT
Telephone	01524592807
Fax	
Mobile	
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	Title	Forename/Initials	Surname
Post	Dr	Fiona	Eccles
Qualifications	MPhys, DPhil, Grad Dip Psych, DClinPsy, Certificate in Academic Practice		
Employer	Lancaster University		
Work Address	Doctorate in Clinical Psychology Furness College, Lancaster University Lancaster		
Post Code	LA1 4YT		
Telephone	01524592807		
Fax			
Mobile			
Work Email	f.eccles@lancaster.ac.uk		
	Title	Forename/Initials	Surname
Post			
Qualifications			
Employer			
Work Address			
Post Code			
Telephone			
Fax			
Mobile			
Work Email			

A64. Details of research sponsor(s)

A64-1. Sponsor

Lead Sponsor

Status:  NHS or HSC care organisation      Commercial status:

Academic

Pharmaceutical industry

Medical device industry

Local Authority

Other social care provider (including voluntary sector or private organisation)

Other

*If Other, please specify:*

Contact person

Name of organisation Lancaster University

Given name Debbie

Family name Knight

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Address                    Research Support Office, B58 Bowland Main, Lancaster University  
 Town/city                Lancaster  
 Post code                LA1 4YT  
 Country                   UNITED KINGDOM  
 Telephone                01524592605  
 Fax  
 E-mail                    ethics@lancaster.ac.uk

Is the sponsor based outside the UK?

 Yes    No

*Under the Research Governance Framework for Health and Social Care, a sponsor outside the UK must appoint a legal representative established in the UK. Please consult the guidance notes.*

A65. Has external funding for the research been secured?

- Funding secured from one or more funders  
 External funding application to one or more funders in progress  
 No application for external funding will be made

What type of research project is this?

- Standalone project  
 Project that is part of a programme grant  
 Project that is part of a Centre grant  
 Project that is part of a fellowship/ personal award/ research training award  
 Other

Other – please state :

A67. Has this or a similar application been previously rejected by a Research Ethics Committee in the UK or another country?

 Yes    No

*Please provide a copy of the unfavourable opinion letter(s). You should explain in your answer to question A6-2 how the reasons for the unfavourable opinion have been addressed in this application.*

A69-1. How long do you expect the study to last in the UK?

Planned start date: 03.08.2015

Planned end date: 13.05.2016

Total duration:

Years: 0 Months: 9 Days: 11

A71-2. Where will the research take place? (Tick as appropriate)

- England  
 Scotland

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- Wales  
 Northern Ireland  
 Other countries in European Economic Area

Total UK sites in study

Does this trial involve countries outside the EU?

- Yes  No

A72. What host organisations (NHS or other) in the UK will be responsible for the research sites? Please indicate the type of organisation by ticking the box and give approximate numbers of planned research sites:

- NHS organisations in England  
 NHS organisations in Wales  
 NHS organisations in Scotland  
 HSC organisations in Northern Ireland  
 GP practices in England  
 GP practices in Wales  
 GP practices in Scotland  
 GP practices in Northern Ireland  
  
 Phase 1 trial units  
 Prison establishments  
 Probation areas  
 Independent hospitals  
 Educational establishments 1  
 Independent research units  
 Other (give details)

Total UK sites in study: 1

A76. Insurance/indemnity to meet potential legal liabilities

*Note: in this question to NHS indemnity schemes include equivalent schemes provided by Health and Social Care (HSC) in Northern Ireland*

A76-1. What arrangements will be made for insurance and/or indemnity to meet the potential legal liability of the sponsor(s) for harm to participants arising from the management of the research? Please tick box(es) as applicable.

*Note: Where a NHS organisation has agreed to act as sponsor or co-sponsor, indemnity is provided through NHS schemes. Indicate if this applies (there is no need to provide documentary evidence). For all other sponsors, please describe the arrangements and provide evidence.*

- NHS indemnity scheme will apply (NHS sponsors only)  
 Other insurance or indemnity arrangements will apply (give details below)

Lancaster University legal liability cover will apply

Please enclose a copy of relevant documents.

A76-2. What arrangements will be made for insurance and/or indemnity to meet the potential legal liability of the sponsor(s) or employer(s) for harm to participants arising from the design of the research? Please tick box(es) as

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*applicable.**Note: Where researchers with substantive NHS employment contracts have designed the research, indemnity is provided through NHS schemes. Indicate if this applies (there is no need to provide documentary evidence). For other protocol authors (e.g. company employees, university members), please describe the arrangements and provide evidence.*

- NHS indemnity scheme will apply (protocol authors with NHS contracts only)
- Other insurance or indemnity arrangements will apply (give details below)

Lancaster University legal liability cover will apply

*Please enclose a copy of relevant documents.*A76-3. What arrangements will be made for insurance and/or indemnity to meet the potential legal liability of investigators/collaborators arising from harm to participants in the conduct of the research?*Note: Where the participants are NHS patients, indemnity is provided through the NHS schemes or through professional indemnity. Indicate if this applies to the whole study (there is no need to provide documentary evidence). Where non-NHS sites are to be included in the research, including private practices, please describe the arrangements which will be made at these sites and provide evidence.*

- NHS indemnity scheme or professional indemnity will apply (participants recruited at NHS sites only)
- Research includes non-NHS sites (give details of insurance/indemnity arrangements for these sites below)

Lancaster University legal liability cover will apply

*Please enclose a copy of relevant documents.*

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**PART C: Overview of research sites**

Please enter details of the host organisations (Local Authority, NHS or other) in the UK that will be responsible for the research sites. For NHS sites, the host organisation is the Trust or Health Board. Where the research site is a primary care site, e.g. GP practice, please insert the host organisation (PCT or Health Board) in the Institution row and insert the research site (e.g. GP practice) in the Department row.

Research site		Investigator/ Collaborator/ Contact	
Institution name	Lancaster University	Title	Miss
Department name	Doctorate in Clinical Psychology, Division of Health Research	First name/ Initials	Rachael
Street address	Furness College, Lancaster University	Surname	Theed
Town/city	Lancaster		
Post Code	LA1 4YT		
Participant Identification Centre(PIC)-Collaborator/ Contact			
PIC contact is [REDACTED]			

Date: 27/07/2015

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**PART D: Declarations****D1. Declaration by Chief Investigator**

1. The information in this form is accurate to the best of my knowledge and belief and I take full responsibility for it.
2. I undertake to abide by the ethical principles underlying the Declaration of Helsinki and good practice guidelines on the proper conduct of research.
3. If the research is approved I undertake to adhere to the study protocol, the terms of the full application as approved and any conditions set out by review bodies in giving approval.
4. I undertake to notify review bodies of substantial amendments to the protocol or the terms of the approved application, and to seek a favourable opinion from the main REC before implementing the amendment.
5. I undertake to submit annual progress reports setting out the progress of the research, as required by review bodies.
6. I am aware of my responsibility to be up to date and comply with the requirements of the law and relevant guidelines relating to security and confidentiality of patient or other personal data, including the need to register when necessary with the appropriate Data Protection Officer. I understand that I am not permitted to disclose identifiable data to third parties unless the disclosure has the consent of the data subject or, in the case of patient data in England and Wales, the disclosure is covered by the terms of an approval under Section 251 of the NHS Act 2006.
7. I understand that research records/data may be subject to inspection by review bodies for audit purposes if required.
8. I understand that any personal data in this application will be held by review bodies and their operational managers and that this will be managed according to the principles established in the Data Protection Act 1998.
9. I understand that the information contained in this application, any supporting documentation and all correspondence with review bodies or their operational managers relating to the application :
  - Will be held by the REC (where applicable) until at least 3 years after the end of the study; and by NHS R&D offices (where the research requires NHS management permission) in accordance with the NHS Code of Practice on Records Management.
  - May be disclosed to the operational managers of review bodies, or the appointing authority for the REC (where applicable), in order to check that the application has been processed correctly or to investigate any complaint.
  - May be seen by auditors appointed to undertake accreditation of RECs (where applicable).
  - Will be subject to the provisions of the Freedom of Information Acts and may be disclosed in response to requests made under the Acts except where statutory exemptions apply.
  - May be sent by email to REC members.
10. I understand that information relating to this research, including the contact details on this application, may be held on national research information systems, and that this will be managed according to the principles established in the Data Protection Act 1998.
11. Where the research is reviewed by a REC within the UK Health Departments Research Ethics Service, I understand that the summary of this study will be published on the website of the National Research Ethics Service (NRES), together with the contact point for enquiries named below. Publication will take place no earlier than 3 months after issue of the ethics committee's final opinion or the withdrawal of the application.

Contact point for publication (*Not applicable for R&D Forms*)

*NRES would like to include a contact point with the published summary of the study for those wishing to seek further information. We would be grateful if you would indicate one of the contact points below.*

- Chief Investigator  
 Sponsor

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- Study co-ordinator
- Student
- Other – please give details
- None

Access to application for training purposes (*Not applicable for R&D Forms*)

*Optional – please tick as appropriate:*

I would be content for members of other RECs to have access to the information in the application in confidence for training purposes. All personal identifiers and references to sponsors, funders and research units would be removed.

This section was signed electronically by Miss Rachael Theed on 23/07/2015 15:59.

Job Title/Post: Trainee Clinical Psychologist  
Organisation: Lancaster University  
Email: r.theed@lancaster.ac.uk

Date: 27/07/2015

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**D2. Declaration by the sponsor's representative**

*If there is more than one sponsor, this declaration should be signed on behalf of the co-sponsors by a representative of the lead sponsor named at A64-1.*

I confirm that:

1. This research proposal has been discussed with the Chief Investigator and agreement in principle to sponsor the research is in place.
2. An appropriate process of scientific critique has demonstrated that this research proposal is worthwhile and of high scientific quality.
3. Any necessary indemnity or insurance arrangements, as described in question A76, will be in place before this research starts. Insurance or indemnity policies will be renewed for the duration of the study where necessary.
4. Arrangements will be in place before the study starts for the research team to access resources and support to deliver the research as proposed.
5. Arrangements to allocate responsibilities for the management, monitoring and reporting of the research will be in place before the research starts.

Please note: *The declarations below do not form part of the application for approval above. They will not be considered by the Research Ethics Committee.*

6. Where the research is reviewed by a REC within the UK Health Departments Research Ethics Service, I understand that the summary of this study will be published on the website of the National Research Ethics Service (NRES), together with the contact point for enquiries named in this application. Publication will take place no earlier than 3 months after issue of the ethics committee's final opinion or the withdrawal of the application.
7. Specifically, for submissions to the Research Ethics Committees (RECs) I declare that any and all clinical trials approved by the HRA since 30th September 2013 (as defined on IRAS categories as clinical trials of medicines, devices, combination of medicines and devices or other clinical trials) have been registered on a publicly accessible register in compliance with the HRA registration requirements for the UK, or that any deferral granted by the HRA still applies.

This section was signed electronically by An authorised approver at ethics@lancaster.ac.uk on 24/07/2015 09:19.

Job Title/Post: Research Support Officer  
 Organisation: Lancaster University  
 Email: s.c.taylor@lancaster.ac.uk

Date: 27/07/2015

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## D8. Declaration for student projects by academic supervisor(s)

1. I have read and approved both the research proposal and this application. I am satisfied that the scientific content of the research is satisfactory for an educational qualification at this level.
2. I undertake to fulfil the responsibilities of the supervisor for this study as set out in the Research Governance Framework for Health and Social Care.
3. I take responsibility for ensuring that this study is conducted in accordance with the ethical principles underlying the Declaration of Helsinki and good practice guidelines on the proper conduct of research, in conjunction with clinical supervisors as appropriate.
4. I take responsibility for ensuring that the applicant is up to date and complies with the requirements of the law and relevant guidelines relating to security and confidentiality of patient and other personal data, in conjunction with clinical supervisors as appropriate.

## Academic supervisor 1

This section was signed electronically by Dr Fiona Eccles on 23/07/2015 16:06.

Job Title/Post: Lecturer in Health Research  
 Organisation: Lancaster University  
 Email: feccles@lancaster.ac.uk

## Academic supervisor 2

This section was signed electronically by jane simpson on 27/07/2015 10:14.

Job Title/Post: Research Director  
 Organisation: Lancaster University  
 Email: j.simpson2@lancaster.ac.uk

Date: 27/07/2015

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## Health Research Authority

18 September 2015

Miss Rachael Theed  
Furness College  
Lancaster University  
Lancaster  
LA1 4YT

Dear Miss Theed

**Study title:** Understandings of psychological difficulties in Huntington's disease and expectations of psychological therapy

**REC reference:** 15/YH/0377  
**IRAS project ID:** 184010

Thank you for your letter of 14 September 2015, responding to the Committee's request for further information on the above research and submitting revised documentation.

The further information has been considered on behalf of the Committee by the Chair.

We plan to publish your research summary wording for the above study on the HRA website, together with your contact details. Publication will be no earlier than three months from the date of this opinion letter. Should you wish to provide a substitute contact point, require further information, or wish to make a request to postpone publication, please contact the REC Manager, [REDACTED], [REDACTED].

### Confirmation of ethical opinion

On behalf of the Committee, I am pleased to confirm a **Favourable** ethical opinion for the above research on the basis described in the application form, protocol and supporting documentation as revised, subject to the conditions specified below.

### Conditions of the favourable opinion

The favourable opinion is subject to the following conditions being met prior to the start of the study.

The Chair noted that if recruitment is expanded and an additional site included then this would need to be notified to the REC via submission of a substantial amendment.

Management permission or approval must be obtained from each host organisation prior to the start of the study at the site concerned.

*Management permission ("R&D approval") should be sought from all NHS organisations*

*involved in the study in accordance with NHS research governance arrangements.*

Guidance on applying for NHS permission for research is available in the Integrated Research Application System or at <http://www.rdforum.nhs.uk>.

*Where a NHS organisation's role in the study is limited to identifying and referring potential participants to research sites ("participant identification centre"), guidance should be sought from the R&D office on the information it requires to give permission for this activity.*

*For non-NHS sites, site management permission should be obtained in accordance with the procedures of the relevant host organisation.*

*Sponsors are not required to notify the Committee of approvals from host organisations*

### Registration of Clinical Trials

All clinical trials (defined as the first four categories on the IRAS filter page) must be registered on a publically accessible database within 6 weeks of recruitment of the first participant (for medical device studies, within the timeline determined by the current registration and publication trees).

There is no requirement to separately notify the REC but you should do so at the earliest opportunity e.g. when submitting an amendment. We will audit the registration details as part of the annual progress reporting process.

To ensure transparency in research, we strongly recommend that all research is registered but for non-clinical trials this is not currently mandatory.

If a sponsor wishes to contest the need for registration they should contact Catherine Blewett ([catherineblewett@nhs.net](mailto:catherineblewett@nhs.net)), the HRA does not, however, expect exceptions to be made. Guidance on where to register is provided within IRAS.

**It is the responsibility of the sponsor to ensure that all the conditions are complied with before the start of the study or its initiation at a particular site (as applicable).**

### **Ethical review of research sites**

#### NHS sites

The favourable opinion applies to all NHS sites taking part in the study, subject to management permission being obtained from the NHS/HSC R&D office prior to the start of the study (see "Conditions of the favourable opinion" below).

### **Approved documents**

The final list of documents reviewed and approved by the Committee is as follows:

<i>Document</i>	<i>Version</i>	<i>Date</i>
Copies of advertisement materials for research participants [Appendix 5: Cover letter]	2	13 September 2015
Evidence of Sponsor insurance or indemnity (non NHS Sponsors only) [Professional Indemnity]	2	13 September 2015
Interview schedules or topic guides for participants [Appendix 1: Interview Schedule (PwHD)]	2	13 September 2015
Interview schedules or topic guides for participants [Appendix 2: Interview Schedule (Partners)]	2	13 September 2015
Letter from sponsor [Sponsor Letter]	1	23 July 2015

Other [Supervisor CV]	1	29 July 2015
Other [Review Letter]	1	03 August 2015
Other [Appendix 8: Debrief Sheet]	2	13 September 2015
Other [Insurance: Employers' Liability]	2	13 September 2015
Other [Insurance: Public Liability]	2	13 September 2015
Other [Response Letter]	1	14 September 2015
Participant consent form [Appendix 4: Consent to contact form]	2	13 September 2015
Participant consent form [Appendix 6: Consent form (PwHD)]	2	13 September 2015
Participant consent form [Appendix 7: Consent form (Partners)]	2	13 September 2015
Participant information sheet (PIS) [Appendix 3: Participant Information Sheet]	2	13 September 2015
REC Application Form [REC_Form_14092015]		14 September 2015
Research protocol or project proposal [Research Protocol]	2	13 September 2015
Summary CV for Chief Investigator (CI) [CV]	1	17 July 2015
Summary CV for supervisor (student research) [Supervisor CV]	1	17 July 2015

### Statement of compliance

The Committee is constituted in accordance with the Governance Arrangements for Research Ethics Committees and complies fully with the Standard Operating Procedures for Research Ethics Committees in the UK.

### After ethical review

#### Reporting requirements

The attached document "*After ethical review – guidance for researchers*" gives detailed guidance on reporting requirements for studies with a favourable opinion, including:

- Notifying substantial amendments
- Adding new sites and investigators
- Notification of serious breaches of the protocol
- Progress and safety reports
- Notifying the end of the study

The HRA website also provides guidance on these topics, which is updated in the light of changes in reporting requirements or procedures.

### User Feedback

The Health Research Authority is continually striving to provide a high quality service to all applicants and sponsors. You are invited to give your view of the service you have received and the application procedure. If you wish to make your views known please use the feedback form available on the HRA website: <http://www.hra.nhs.uk/about-the-hra/governance/quality-assurance/>

### HRA Training

We are pleased to welcome researchers and R&D staff at our training days – see details at <http://www.hra.nhs.uk/hra-training/>

<b>15/YH/0377</b>	<b>Please quote this number on all correspondence</b>
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With the Committee's best wishes for the success of this project.

Yours sincerely



pp [REDACTED]

**Chair**

Email: [REDACTED]

*Enclosures:* 'After ethical review – guidance for researchers'

*Copy to:* Ms Debbie Knight – R&D Dept, Lancaster University



NHS Foundation Trust

[Redacted text block]

Miss Rachael Theed  
Furness College  
Lancaster University  
Lancaster  
LA1 4YT

Date: 02 October 2015

Our Ref: [Redacted]

Dear Miss Theed,

Study: Understandings of psychological difficulties in Huntington’s disease and expectations of psychological therapy  
PIN: [Redacted]  
REC reference: 15/YH/0377  
Sponsor: Lancaster University  
Chief Investigator: Miss Rachael Theed  
Local Liaison: [Redacted]

We have received a request for authorisation for our Trust to become involved as a **Participant Identification Centre (PIC)** for the above study.

Following receipt of the documentation listed at the foot of this letter, we have completed the checks required for a PIC site and can confirm our agreement.

I would like to take this opportunity to wish you well with your research.

Yours sincerely

[Redacted]  
**Research Support Manager**

CC: [Redacted]  
Academic Supervisors – Dr Jane Simpson & Dr Fiona Eccles

Documents Acknowledged	Version Number / Reference	Date
REC Favourable Opinion Letter		18 September 2015
Appendix 5: Cover Letter	2	13 September 2015

## R04122 R&amp;D Approval -PIC

Professional Indemnity		
Appendix 1: Interview Schedule PwHD	2	13 September 2015
Appendix 2: Interview Schedule Partners	2	13 September 2015
Sponsor Letter		21 July 2015
Supervisor CV – F. Eccles		
Review Letter		03 August 2015
Appendix 8: Debrief Sheet	2	13 September 2015
Insurance: Employers Liability		
Insurance: Public Liability		
Response Letter		
Appendix 4: Consent to Contact Form	2	13 September 2015
Appendix 6: Consent Form (PwHD)	2	13 September 2015
Appendix 7: Consent Form (partners)	2	13 September 2015
Appendix 3: Participant Information Sheet	2	13 September 2015
Research Protocol	2	13 September 2015
CV for CI – R. Theed		
Supervisor CV – J. Simpson		

**Appendices**

Rachael Theed

Trainee Clinical Psychologist

Doctorate in Clinical Psychology

Division of Health Research, Lancaster University

Word Count

**Appendix 4-1: Research Protocol**



Thesis: Research Protocol

Understandings of psychological difficulties in Huntington's disease and expectations of psychological therapy

Rachael Theed

Trainee Clinical Psychologist

Lancaster University

Supervised by Dr Jane Simpson, Research Director, Lancaster University and

Dr Fiona Eccles, Lecturer in Research Methods, Lancaster University

## Introduction

Huntington's disease (HD) is a neurodegenerative condition causing problems with cognitive functioning, coordination, movement and some emotional difficulties. It is suggested that around five to ten per 100,000 people are affected (Kay, Fisher & Hayden, 2014). People are generally diagnosed between the ages of 35-55 with a life expectancy of around 15-20 years (Keenan, Simpson, Miedzybrodzka, Alexander & Semper, 2013) and have often seen their parents affected by the disease (Kremer, 2002).

People with HD often experience emotional difficulties including depression, anxiety, apathy and irritability (Kirkwood, Su, Conneally & Foroud, 2001) which have the potential to impact on an individual's quality of life. For example, rates of depression have been shown to be high in individuals with HD with the causes for this being potentially twofold (Paulsen et al., 2005). Certainly one potential cause is having to adjust to living with the illness as well as coping with the subsequent difficulties that arise as a result of HD. Additionally, there is a potential biological component whereby neural mechanisms in the brain are affected by HD subsequently affecting mood (Paulsen et al., 2005).

Importantly, there are significant physical and emotional consequences of HD not only for the individual with the disease but also for the people with whom they live (e.g., Aubeeluck, Buchanan & Stupple, 2012; Williams et al., 2009). Family members often take on caregiving for the individual with HD and subsequently may experience a reduced quality of life, including lowered mood (Aubeeluck & Buchanan, 2007; McCabe, Firth & O'Connor, 2009; Read et al., 2010), potentially as a result of witnessing a loved one become increasingly unwell over time and grieving for a lost relationship (Pickett, Altmaier & Paulsen, 2007). Indeed, depression in people with HD has been identified as being a strong predictor of depression in those caring for someone with HD (Pickett et al., 2007).

Although for people living with HD medication is often the main approach in supporting people with mood difficulties, psychological interventions may provide an alternative way to reduce distress. Medication may not always be the preferred option for people with HD and there could be alternative options. For example, in another neurodegenerative disease, Parkinson's disease (PD), while medication may be used to support people with the psychological difficulties they experience, there is increasing evidence to support the use of psychological interventions in this population (Charidimou, Seamons, Selai & Schrag, 2011; Dobkins et al., 2011), including mindfulness-based cognitive therapy (MBCT; Fitzpatrick, Simpson & Smith, 2010). Due to the level of distress both people with HD and their partners experience, it is important to understand what people think about the support available and treatment they receive.

MBCT is an eight week group therapy developed by Segal, Williams and Teasdale (2002) which teaches mindfulness skills through a range of practices with the aim of preventing the reoccurrence of depression (Gu, Strauss, Bond & Cavanagh, 2015). Previously MBCT has been piloted with people with Parkinson's disease who found it improved their psychological well-being (Fitzpatrick, Simpson & Smith, 2010). MBCT is therefore going to be piloted for individuals with HD with the aim of alleviating psychological distress directly in people with HD and indirectly in their partners. Although the partners will not be directly participating in the MBCT groups, higher levels of mindfulness have been shown to be associated with higher levels of satisfaction in partner relationships (Barnes, Brown, Krusemark, Campbell & Rogge, 2007). Furthermore, people engaged in MBCT have reported increased empathy and perspective taking, allowing them to respond more mindfully in relationships (Bihari & Mullan, 2014).

Consequently, this study aims to investigate both individuals with HD and their partners' understanding of psychological difficulties in HD and their views of psychological

therapy. Participants will be recruited from those who are due to take part in the trial of MBCT and therefore the study will focus in particular on mindfulness and people's knowledge of this, as well as their hopes and expectations for MBCT. This study will adopt a qualitative methodology to obtain detailed accounts of people with HD and their partners understanding of psychological difficulties and expectations of psychological therapy. In order to address this, semi-structured interviews will be conducted and analysed by interpretative phenomenological analysis (IPA; Smith, 2009).

## **Method**

### **Participants**

This study will aim to recruit between 3-6 participants with HD and 3-6 partners of a person with HD. However, if one group (either people with HD or their partners) is unable to be recruited then up to 12 participants from the other group may be recruited. Participants will be recruited from the MBCT trial. The person with HD will be signed up to engage in a MBCT programme but does not have to have a partner to take part in the study. Furthermore, if an insufficient number of participants are recruited then an additional site [REDACTED] may be included in the study.

To participate in the present study participants must meet the following criteria:

- People with HD will be signed up to participate in the MBCT trial.
- Partners of those signed up to participate in the MBCT trial.
- Participants must be aged 18 or over.
- Participants must understand and be able to speak English.

People with HD and their partners who are not signed up to engage in the MBCT programme will not be eligible for inclusion.

**Design**

This study will adopt a qualitative design. Semi-structured interviews with people with HD and their partners will be conducted prior to them engaging in the MBCT programme. Interviews will be analysed by IPA (Smith, 2009), however if there is an insufficient number of participants (either in people with HD or their partners) then thematic analysis will be used to analyse the data.

**Materials**

The interviews will be guided by a semi-structured interview schedule (appendix 1 & 2) which will be informed by previous qualitative studies which have looked at the effectiveness of mindfulness-based interventions (Cairns & Murray, 2015) as well as a study conducted with a neurological population (Fitzpatrick et al., 2010). However, further questions may be asked which are sensitive to, and influenced by, participants' responses.

**Procedure**

People with HD will be recruited from the MBCT trial. Potential participants will be informed about the research when they have opted into the MBCT trial. Those suitable for inclusion in the study will be provided with an information pack including a participant information sheet (appendix 3) and consent to contact form (appendix 4) either via post, email or in person by the recruiters for the main MBCT trial (Dr Fiona Eccles & Dr Jane Simpson), with a cover letter (appendix 5). As partners will be recruited through the person with HD engaging in the MBCT programme, if they are interested, an information pack for the partner will be sent to the person with HD via email or post. They will then be asked to pass this on to their partner. The information pack will contain an information sheet and consent to contact form. They will then be asked to contact the researcher (via the consent to

contact form, email or telephone) for an initial discussion regarding the research to consider whether they would like to take part.

Due to the study only requiring a small number of participants there is the potential for people to opt into the study once the target has been reached. In this instance, potential participants will be contacted by the researcher informing them that the target sample has been met.

If people with HD and their partners decide to take part in the research then a mutually convenient interview time will be arranged. Participants will be interviewed individually either at home or at a community location convenient for the participant. People's partners do not have to take part in order for the person with HD to take part and vice versa. However, if the person with HD decides not to participate then partner will still be recruited in the same way via the person with HD. Before commencing the interview the researcher will check each participant has read the participant information sheet and go through the consent form (appendix 6 & 7), answering any questions participants may have. Participants will be interviewed prior to the person with HD engaging in the MBCT programme. Interviews are anticipated to last approximately 60 minutes per person. At the end of the interview participants will be debriefed using the debrief sheet containing sources of support should they require this.

Telephone interviews may also be conducted if participants are not able to meet. If this is the case, a consent form will be posted to the participant and returned prior to the interview. Participants will be debriefed over the phone as well as having the debrief sheet (appendix 8) posted or emailed to them following the interview.

### **Proposed Analysis**

Following each interview, the data will be transcribed by the researcher. IPA will be used to analyse the data, following the stages outlined by Smith (2009). IPA is widely used in psychological research and aims to explore how homogenous groups of people understand and make sense of their personal and social world (Smith, Flowers & Larkin, 2009).

Interviews will be transcribed and analysed individually, in turn. Initial notations relating to the data will be made in the margin which will be used to identify potential themes across transcripts. Data will then be organised into superordinate themes including sub-themes where appropriate.

### **Practical Issues**

The Lancaster University Doctorate in Clinical Psychology's data security and storage policies will be followed. Interviews will be audio recorded then transferred to a computer as soon as possible and stored on the University server via the VPN and password protected. At this point they will be deleted from the device. These recordings will be destroyed once the data has been transcribed, checked and analysed. During the study, transcriptions will also be password protected and stored on the University server. Following submission of the research paper, the data (consent forms and coded data) will be scanned and stored securely for 10 years by the Doctorate in Clinical Psychology's admin team, while the original paper copies will be destroyed. At the end of this period the data will be destroyed. Consent to contact forms will be kept until participants have received a copy of the results should they have requested these.

As the interviews will be conducted on a one to one basis, when lone working the researcher will follow the [REDACTED] lone working policy. See [http://www.lancaster.ac.uk/shm/study/doctoral\\_study/dclinpsy/new/onlinehandbook/appendices/lcft\\_lone\\_working\\_policy.pdf](http://www.lancaster.ac.uk/shm/study/doctoral_study/dclinpsy/new/onlinehandbook/appendices/lcft_lone_working_policy.pdf). A designated person (a fellow trainee) will be given the

researcher's contact details and information about the appointment time and duration. They will further be provided with contact details of the interview location and interviewee (in a sealed envelope, only to be opened in an emergency). When the interview is finished the researcher will contact the designated person to inform them that the interview is complete. If they do not receive this contact then attempts will be made to contact the researcher. If they are unable to make contact then they can open the sealed envelope and the appropriate authorities will be informed.

### **Ethical Concerns**

Ethical approval will be obtained through the National Research Ethics Service (NRES) and research governance approval from [REDACTED] [REDACTED] research and development department who will act as the participant identification centre (PIC). All participant information will remain confidential. The researcher will not have access to potential participants' personal information until the participants themselves express an interest in participating in the study and provide their own contact details. The researcher and their supervisors will be the only people to have access to the audio recordings and transcripts. This will be outlined on the participants information sheet and consent form.

However, there may be the potential for participants to disclose information that highlights a potential risk of harm, either to themselves or others. If issues related to risk are disclosed then confidentiality may need to be broken and the information disclosed with the appropriate individuals. Again, participants will be informed of this exception to confidentiality prior to commencing the interview.

While it is not expected that the research will cause participants any distress, if this occurs during interviews the participant will be given a break and asked if they would like to

continue. They will also be full debriefed at the end of the interview and provided with a list of contacts for support.

**Timescale**

June - July 2015: Submit to ethics process.

August – December 2015: Recruitment, data collection and analysis

October 2015: Draft read of Introduction and Method sections.

January – April 2016: Write up and submit drafts.

May 2016: Submit research paper.

### References

- Aubeeluck, A., Buchanan, H. & Stupple, E. N. (2012). 'All the burden on all the carers': exploring quality of life with family caregivers of Huntington's disease patients. *Quality of Life Research*, 21, 1425-1435. doi: 10.1007/s11136-011-0062-x.
- Aubeeluck, A. & Buchanan, H. (2007). The Huntington's Disease quality of life battery for carers: reliability and validity. *Clinical Genetics*, 71, 434-445. doi: 10.1111/j.1399-0004.2007.00784.x.
- Barnes, S., Brown, K. W., Krusemark, E., Campbell, W. K. & Rogge, R. D. (2007). The role of mindfulness in romantic relationship satisfaction and responses to relationship stress. *Journal of Marital and Family Therapy*, 33 (4), 482-500. doi: 10.1111/j.1752-0606.2007.00033.x.
- Bihari, J. & Mullan, E. (2014). Relating Mindfully: A Qualitative Exploration of Changes in Relationships Through Mindfulness-Based Cognitive Therapy. *Mindfulness*, 5, 46-59. doi: 10.1007/s12671-012-0146-x.
- Cairns, V., & Murray, C. (2015). How do the features of Mindfulness-Based Cognitive Therapy contribute to positive therapeutic change? A meta-synthesis of qualitative studies. *Behavioural and Cognitive Psychotherapy*, 43 (3), 342-359. doi: 10.1017/S1352465813000945.
- Charidimou, A., Seamons, J., Selai, C., & Schrag, A. (2011). The role of Cognitive-Behavioural Therapy for patients with depression in Parkinson's disease. *Parkinson's disease*, 2011. doi: 10.4061/2011/737523.
- Dobkin, R.D., Menza, M., Allen, L.A., Gara, M.A., Mark, M.H., Tiu, J., ... Friedman, J. (2011). Cognitive behavioural therapy for depression in Parkinson's disease: A randomised, controlled trial. *American Journal of Psychiatry*, 168 (10), 1066-1074. doi: 10.1176/appi.ajp.2011.10111669.

- Fitzpatrick, L., Simpson, J. & Smith, A. (2010). A qualitative analysis of mindfulness-based cognitive therapy (MBCT) in Parkinson's disease. *Psychology and Psychotherapy: Theory, Research and Practice*, 83 (2), 179-192. doi: 10.1348/147608309X471514.
- Gu, J., Strauss, C., Bond., R., & Cavanagh, K. (2015). How do mindfulness-based cognitive therapy and mindfulness-based stress reduction improve mental health and well-being? A systematic review and meta-analysis of mediation studies. *Clinical Psychology Review*, 37, 1-12. doi: 10.1016/j.cpr.2015.01.006.
- Kay, C., Fisher, E. & Hayden, M. R. 2014. Epidemiology. In: Bates, G., Tabrizi, S. J. & Jones, L. (eds.) *Huntington's disease*. 4th ed. Oxford: Oxford University Press.
- Keenan, K.F., Simpson, S.A., Miedzybrodzka, Z., Alexander, D.A., & Semper, J. (2013). How do partners find out about the risk of Huntington's disease in couple relationships. *Journal of Genetic Counselling*, 22, 336-344. doi: 10.1007/s10897-012-9562-2.
- Kirkwood, S.C., Su, J.L., Conneally, M., & Foroud, T. (2001). Progression of symptoms in the early and middle stages of Huntington's disease. *Archives of Neurology*, 58 (2), 273-278. doi: 10.1001/archneur.58.2.273.
- Kremer, B. 2002. Clinical neurology in Huntington's disease. In: Bates, G., Harper, P. & Jones, L. (eds.) *Huntington's disease*. 3rd ed. Oxford: Oxford University Press.
- Mccabe, M. P., Firth, L. & O'connor, E. (2009). A Comparison of Mood and Quality of Life Among People with Progressive Neurological Illnesses and Their Caregivers. *Journal of Clinical Psychology in Medical Settings*, 16, 355-362. doi: 10.1007/s10880-009-9168-5.

- Paulsen, J.S., Nehl, C., Hoth, K.F., Kanz, J.E., Benjamin, M., Conybeare, R., ... & Turner, B. (2005). Depression and stages of Huntington's disease. *Journal of Neuropsychiatry and Clinical Neurosciences*, 17 (4), 496-502. doi: 10.1176/jnp.17.4.496.
- Pickett, T., Altmaier, E., & Paulsen, J.S. (2007). Caregiver burden in Huntington's disease. *Rehabilitation Psychology*, 52 (3), 311-318. doi: 10.1037/0090-5550.52.3.311.
- Read, J. E., A, T. P., Langbehn, D. R., Owen, G., Dürr, A., Leavitt, B. R., Roos, R. a. C., Stout, J. C., Tabrizi, S. J., Craufurd, D. & Investigators, T.-H. (2010). Quality of life in Huntington's disease: a comparative study investigating the impact on spouses of those with premanifest and early disease. *Journal of Neurology, Neurosurgery & Psychiatry*, 81, A23-A24. doi: 10.1136/jnnp.2010.222620.4.
- Segal, Z.V., Williams, J.M., & Teasdale, J. (2002). *Mindfulness-based cognitive therapy for depression: A new approach to preventing relapse*. London, UK: Guildford Press.
- Smith, J. A. (2009). *Interpretative phenomenological analysis : theory, method and research*. London, Sage.
- Smith, J.A., Flowers, P. & Larkin, M. (2009). *Interpretative phenomenological analysis: theory method and research: understanding method and application*. London, Sage.
- Williams, J. K., Skirton, H., Paulsen, J. S., Tripp-Reimer, T., Jarmon, L., Mcgonigal Kenney, M., Birrer, E., Hennig, B. L. & Honeyford, J. (2009). The emotional experiences of family carers in Huntington disease. *Journal of Advanced Nursing*, 65, 789-798. doi: 10.1111/j.1365-2648.2008.04946.x.

## Appendix 4-2: Interview Schedule



### **Understandings of psychological difficulties in Huntington's disease and expectations of psychological therapy**

#### **Semi-structured interview schedule: People with Huntington's disease**

This interview schedule gives an indication of the topic areas to be discussed in the interview with example questions. The precise questions will be dependent on participants' responses and the focus of each interview will be guided in part by what is deemed important to the individual being interviewed.

This interview will be conducted prior to the start of the MBCT programme. The interviewer will explore the psychological therapy experiences of the participant prior to the start of the MBCT course. It will include their emotional and psychological wellbeing prior to either themselves or their partner engaging in the course as well as their expectations of the course.

Example questions:

#### **Introduction**

For how long have you known that you have had the HD gene?

Do you think you show any signs of the condition at the moment?

Do you think having HD affects how you feel? Does this also affect your partner?

If you have thought about how you feel, what is your understanding of psychological distress and where this comes from?

#### **Therapy expectations and experiences**

If you have ever thought about psychological therapy, do you think it could be helpful for people living with HD and their partners? What makes you think that?

Have you had any previous experience of psychological therapy?

If you have had experience, what did you find beneficial about any previous therapy you have received?

Have you experienced anything similar to MBCT in the past? (i.e. mindfulness)

When you were first approached about engaging in the MBCT trial what were your first thoughts about the course?

What led to your decision to take part in the research? How were you feeling in yourself when you decided to take part? How was your partner feeling?

Do you or your partner have any expectations as to what the course will be like or the impact it may have on you or your partner?

Do you expect the course to help you and/or your partner? In what way do you think it might help?

Do you have any worries about the MBCT course?

Do you think there will be an impact on your psychological and emotional wellbeing? If so, what?

Do you think there will be a wider impact on you and/or your partner? (e.g. your relationship and/or wider family relationships).

**Conclusion**

Is there anything else you think it would be useful for us to know?

Thanks and debrief.

**Appendix 4-3: Interview Schedule****Understandings of psychological difficulties in Huntington's disease and expectations of psychological therapy****Semi-structured interview schedule: Partners of a person with Huntington's disease**

This interview schedule gives an indication of the topic areas to be discussed in the interview with example questions. The precise questions will be dependent on participants' responses and the focus of each interview will be guided in part by what is deemed important to the individual being interviewed.

This interview will be conducted prior to the start of the MBCT programme. The interviewer will explore the psychological therapy experiences of the participant prior to the start of the MBCT course. It will include their emotional and psychological wellbeing prior to either themselves or their partner engaging in the course as well as their expectations of the course.

Example questions:

**Introduction**

For how long have you known that your partner has had the HD gene?

Do you think he/she shows any signs of the condition at the moment?

Do you think your partner having HD affects how you feel? Does this also affect your partner?

If you have thought about how you feel, what is your understanding of psychological distress and where this comes from?

**Therapy expectations and experiences**

If you have ever thought about psychological therapy, do you think it could be helpful for people living with HD and their partners? What makes you think that?

Have you had any previous experiences of psychological therapy?

If you have, what did you find beneficial about any previous therapy you have received?

Have you experienced anything similar to MBCT in the past? (i.e. mindfulness)

Has your partner ever had any psychological therapy in the past? If so, did they find it helpful?

When your partner was approached about engaging in the MBCT trial what were your first thoughts about the course? What did your partner think?

What do you think led to their decision to take part in the research? How was your partner feeling when they decided to take part?

Do you or your partner have any expectations as to what the course will be like or the impact it may have on you or your partner?

Do you expect the course to help your partner? In what way do you think it might help?

Do you have any worries about the MBCT course?

Do you think there will be an impact on both your partners and your psychological and emotional wellbeing? If so, what?

Do you think there will be a wider impact on you and/or your partner? (e.g. your relationship and/or wider family relationships).

**Conclusion**

Is there anything else you think it would be useful for us to know?

Thanks and debrief.

**Appendix 4-4: Participant Information Sheet****Participant Information Sheet****Understandings of psychological difficulties in Huntington's disease and expectations of psychological therapy**

We would like to invite you to take part in a research study, which is being conducted as part of a Doctorate in Clinical Psychology. Before you decide if you would like to take part or not, we would like you to understand why the research is being done and what it would involve for you. We will also go through the information sheet with you and answer any questions you have before you decide whether you want to take part.

**What is the study about?**

People with the gene for Huntington's disease often experience psychological difficulties such as low mood (depression), anxiety and irritability. Usually they are given medication to help with these problems however medication may only help to a certain extent or alternatively some people do not want to take it. Mindfulness-based cognitive therapy (MBCT) is a type of psychological therapy which has been shown to help people with depression and other psychological difficulties and is shortly going to be trialled with people with the gene for Huntington's disease to see if it can help them. The aim of this study is to understand your experiences of any previous psychological therapy and your hopes and expectations of the MBCT trial that either yourself or your partner will be taking part in.

**Why have I been approached?**

Either you or your partner has agreed to be part of the MBCT pilot trial. We would like to understand what people's previous experience and understanding of psychological therapy is prior to engaging in the MBCT trial. We would also like to gain insight in what people (both the individual and their partner) hope to gain from psychological therapy, in particular MBCT.

While partners of people with HD will not be taking part in the MBCT trial, we are interested in their hopes and expectations of the course and how they think it will impact on both their and their partner's life.

**Do I have to take part?**

No. It's completely up to you to decide whether or not you take part. If you decide not to take part it will not affect your clinical care or that of your partner. If you agree to take part, you can stop and withdraw at any time without giving a reason.

**What will I be asked to do if I take part?***Opting in*

If you are interested in taking part then first you need to contact myself (more detail at the end of this information sheet) and I will tell you more about the research. If you are still interested in taking part, I will come and meet with you (either at home or another location near you) so you can sign a consent form.

### *Collecting information*

If you decide to participate we will collect some data from you prior to the course commencing. We would like to do this by conducting interviews. We would like to interview you so you can tell us about any previous experiences of therapy, what impact you think therapy will have both on yourself and your partner and how you think it will impact on your emotional and psychological wellbeing. The interview would last approximately an hour. You can stop the interview at any time and it can be done in two parts if you feel tired. The interview will be audio-recorded and then transcribed (turned into a written transcript). Interviews can be done either at your home or another location convenient for you. If you are not able to meet then they can also be done on the phone.

### **Will my data be confidential?**

The information you provide will be kept confidential. The data collected for this study will be stored securely and only the researchers conducting this study will have access to this data:

- The typed version of your interview will be made anonymous by removing any identifying information including your name. Anonymised direct quotations from your interview may be used in the reports or publications from the study, so your name will not be attached to them.
- Audio recordings will be destroyed and/or deleted after they have been transcribed and checked
- The files on the computer will be encrypted (that is no-one other than the researchers will be able to access them) and the computer itself password protected.
- At the end of the study, written transcripts and consent forms will also be kept securely on the computer for ten years.

There are some limits to confidentiality. If at any point during the interview you say something that makes us think that either you or someone else is at significant risk of harm, we will have to break confidentiality and speak to the person with Huntington's disease's treating clinician (Dr ..... ) or a member of the research team. If possible, we will tell you if we have to do this.

### **What will happen if I decide to leave part way through?**

You can choose to stop participating in the study at any time. If you leave the study up to 3 weeks following the interview then your data (audio recordings and transcripts) will be destroyed and not used in the research. If you leave the study after this time, then the data may remain in the study. However, if you ask us to withdraw your data at any point, every effort will be made to do so up to the point of submission of thesis, but it may not be possible if your data have already been analysed.

### **What will happen to the results?**

The results will be summarised and reported at local groups and will be submitted for publication in academic and/or professional journals. If you would like a copy of the results, please ask the researchers.

### **Are there any risks?**

It is not anticipated that participating in this research will cause distress. However, talking about your thoughts and feelings in an interview can sometimes be upsetting. If during the

research interview you experience any distress, you are advised to inform the researcher and/or contact the resources at the end of this sheet.

**Are there any benefits to taking part?**

There are no known benefits of taking part in this research. However, we hope that it will help us better understand the psychological therapy experiences of people living with HD and their partners. We also hope it will provide some insight into the psychological therapy needs of people with HD and their partners and how this could impact on their emotional and psychological wellbeing.

**Who has reviewed the project?**

This study has been approved by the NHS Research Ethics Service and Central Manchester University Hospitals NHS Foundation Trust Research and Development department.

**Where can I obtain further information? How do I opt in?**

If you might be interested in participating in the study, please contact a member of the research team. You can do this by email to Rachael Theed: [r.theed@lancaster.ac.uk](mailto:r.theed@lancaster.ac.uk) or Dr Fiona Eccles: [f.eccles@lancaster.ac.uk](mailto:f.eccles@lancaster.ac.uk), by telephone (07508406193) or please fill in the contact sheet and send it back in the pre-paid envelope provided.

You will then be provided with more information about the project so you can decide whether you are interested in taking part.

**Complaints**

If you wish to make a complaint or raise concerns about any aspect of this study and do not want to speak to the researchers, you can contact:

Professor Bruce Hollingsworth  
Head of Division of Health Research  
Lancaster University  
Lancaster  
LA1 4YG  
[b.hollingsworth@lancaster.ac.uk](mailto:b.hollingsworth@lancaster.ac.uk)  
01524 594154

**Resources**

It is not anticipated that taking part in this research will cause distress. However, should you feel distressed as a result of taking part you can contact:

Dr Fiona Eccles: [f.eccles@lancaster.ac.uk](mailto:f.eccles@lancaster.ac.uk)

You can also contact your GP.

The following organisations may also provide advice or support.

**Huntington's disease association [www.hda.org.uk](http://www.hda.org.uk)**

There is lots of advice and information on their website. **If you call the head office on 0151 331 5444, they can put you in touch with your regional care advisory service. More information about this service is given here: <http://hda.org.uk/hda/rca>**

**The Samaritans [www.samaritans.org](http://www.samaritans.org)**

The Samaritans offer a non-judgemental listening service. Their phone number is 08457 90 90 90 (charges apply) or you can email them on [jo@samaritans.org](mailto:jo@samaritans.org)

**Appendix 4-5: Consent to Contact Form****Consent to Contact Form****Understandings of psychological difficulties in Huntington's disease and expectations of psychological therapy**

If you are interested in learning more about the study please contact a member of the research team. You can do this by phoning Rachael Theed directly (07508406193), by email ([r.theed@lancaster.ac.uk](mailto:r.theed@lancaster.ac.uk)) or by filling in this form and returning it in the stamped addressed envelope provided and we will then contact you.

Name: \_\_\_\_\_

Name of person who will be participating in the MBCT course:

\_\_\_\_\_

**Contact details**

Telephone number: \_\_\_\_\_

Email address: \_\_\_\_\_

I would prefer to be contacted by (please circle):            phone        email        don't mind

Any other details (e.g. times that are preferable for us to phone you)

\_\_\_\_\_

**Appendix 4-6: Cover Letter**

[To be sent as email or letter, depending on usual method of contact for the participant. Email will come from Dr Fiona Eccles]

Doctorate in Clinical Psychology  
Furness College  
Lancaster University  
Lancaster  
LA1 4YT

Dear .....

You have recently opted into our study of mindfulness-based cognitive therapy (MBCT) for people with the HD gene. We are looking forward to seeing you on the MBCT course in the Autumn.

In the meantime, we have a trainee clinical psychologist, Rachael Theed, who is working with us and is doing a project related to the main MBCT study. She is interested to find out how people with HD and their partners understand the psychological difficulties that people with HD can experience and also what are their hopes for and expectations of the MBCT course. We wondered if you might also be interested in taking part in her study.

The decision to take part or not in Rachael's study will in no way impact on your taking part in the MBCT course. If you do decide to take part, Rachael will arrange to meet you and will take consent separately for this project. More details are found on the attached/enclosed [to be deleted as appropriate] participant information sheet.

If you would like further information then please get in touch with Rachael (details on the participant information sheet). Alternatively you are welcome to contact Fiona for an initial discussion ([f.eccles@lancaster.ac.uk](mailto:f.eccles@lancaster.ac.uk), 01524 592807).

Thank you for taking the time to consider this additional project and we look forward to seeing you soon on the MBCT course.

Yours sincerely,

Jane Simpson (Research Director DClinPsy course, Chief Investigator MBCT study)

Fiona Eccles (Lecturer in Research Methods)

**Appendix 4-7: Consent Form (People living with HD)****Consent Form (People living with HD)****Understandings of psychological difficulties in Huntington's disease and expectations of psychological therapy**

We are asking if you would like to take part in a research study to investigate the views of people living with HD who will be participating in a trial MBCT programme regarding their understanding of psychological difficulties, any previous psychological therapy experiences and hopes for/expectations of the MBCT programme.

Before you consent to participating in the study we ask that you read the participant information sheet and mark each box below with your initials if you agree. If you have any questions or queries before signing the consent form please speak to the principal researcher, Rachael Theed. Contact details are provided on the participant information sheet.

	Please initial the box after each statement
1. I confirm that I have read the participant information sheet and fully understand what is expected of me within this study.	
2. I confirm that I have had the opportunity to ask any questions and to have them answered.	
3. I understand that my participation is voluntary and that I am free to withdraw at any time without giving any reason, without my medical care or legal rights being affected.	
4. I understand that the data collected during the study may be looked at by individuals from Lancaster University, from regulatory authorities or the NHS Trust where it is relevant to my taking part in the research. I give permission for these individuals to have access to this data.	
5. I understand that my interviews will be audio recorded and then made into an anonymised interview transcript.	
6. I understand that the information from my interviews will be pooled with other participants' responses, anonymised and may be published.	
7. I consent to information from the study including quotations from my interviews being used in reports, conferences and training events.	
8. I understand that any information I give will remain strictly confidential to the researchers unless it is thought that there is a	

risk of harm to myself or others, in which case this information may need to be shared with appropriate persons.	
9. I consent to Lancaster University keeping the data from the study for up to 10 years after the study has finished.	
10. I consent to take part in the above study.	

**Name of Participant** \_\_\_\_\_ **Signature** \_\_\_\_\_ **Date**

\_\_\_\_\_

**Name of Researcher** \_\_\_\_\_ **Signature** \_\_\_\_\_

**Date** \_\_\_\_\_

**Appendix 4-8: Consent Form (Partners of a person with HD)****Consent Form (Partners of a person with HD)****Understandings of psychological difficulties in Huntington's disease and expectations of psychological therapy**

We are asking if you would like to take part in a research study to investigate the views of partners of people with HD who will be participating in a trial MBCT programme regarding their understanding of psychological difficulties, any previous psychological therapy experiences and hopes for/expectations of the MBCT programme.

Before you consent to participating in the study we ask that you read the participant information sheet and mark each box below with your initials if you agree. If you have any questions or queries before signing the consent form please speak to the principal researcher, Rachael Theed. Contact details are provided on the participant information sheet.

	Please initial the box after each statement
1. I confirm that I have read the participant information sheet and fully understand what is expected of me within this study.	
2. I confirm that I have had the opportunity to ask any questions and to have them answered.	
3. I understand that my participation is voluntary and that I am free to withdraw at any time without giving any reason, without my medical care or legal rights being affected.	
4. I understand that the data collected during the study may be looked at by individuals from Lancaster University, from regulatory authorities or the NHS Trust where it is relevant to my taking part in the research. I give permission for these individuals to have access to this data.	
5. I understand that my interviews will be audio recorded and then made into an anonymised interview transcript.	
6. I understand that the information from my interviews will be pooled with other participants' responses, anonymised and may be published.	
7. I consent to information from the study including quotations from my interviews being used in reports, conferences and training events.	
8. I understand that any information I give will remain strictly confidential to the researchers unless it is thought that there is a	

risk of harm to myself or others, in which case this information may need to be shared with appropriate persons.	
9. I consent to Lancaster University keeping the data from the study for up to 10 years after the study has finished.	
10. I consent to take part in the above study.	

**Name of Participant** \_\_\_\_\_ **Signature** \_\_\_\_\_ **Date**

\_\_\_\_\_

**Name of Researcher** \_\_\_\_\_ **Signature** \_\_\_\_\_

**Date** \_\_\_\_\_

**Appendix 4-9: Debrief Sheet****Debriefing Sheet****Understandings of psychological difficulties in Huntington's disease and expectations of psychological therapy**

Should you feel you require any support following the interview process you can contact the following:

Rachael Theed (07508406193) or [r.theed@lancaster.ac.uk](mailto:r.theed@lancaster.ac.uk)

Dr Fiona Eccles: [f.eccles@lancaster.ac.uk](mailto:f.eccles@lancaster.ac.uk)

Dr Jane Simpson: [j.simpson2@lancaster.ac.uk](mailto:j.simpson2@lancaster.ac.uk)

You can also contact your GP.

The following organisations may also provide advice or support:

**Huntington's disease association** [www.hda.org.uk](http://www.hda.org.uk)

There is lots of advice and information on their website. **If you call the head office on 0151 331 5444, they can put you in touch with your regional care advisory service. More information about this service is given here:** <http://hda.org.uk/hda/rca>

**The Samaritans** [www.samaritans.org](http://www.samaritans.org)

The Samaritans offer a non-judgemental listening service. Their phone number is 08457 90 90 90 (charges apply) or you can email them on [jo@samaritans.org](mailto:jo@samaritans.org)

**Purpose of the study**

This study is concerned with the psychological therapy experiences of both people with Huntington's disease (HD) and partners of people with HD. Previous studies have shown that both people with HD and their partners may experience emotional and psychological difficulties, for example low mood, anxiety and irritability, when living with HD. However, the treatment of Huntington's disease and the difficulties arise as a result of living with HD

often adopts a medical focus. This study therefore aimed to gain insight into your experiences of and hopes for psychological therapy.

In this study you were asked questions about your experiences of any previous psychological therapy you or your partner may have received and your hopes and expectations of mindfulness-based cognitive therapy (MBCT). All participants were asked similar questions to help us understand your views.

### **Why is this important to study?**

It is important to understand the psychological therapy experiences of both people with HD and their partners to identify what people feel is and would be beneficial to their emotional and psychological wellbeing. It may further provide evidence for the need for those with HD and their partners who may be struggling to be able to access psychological therapy.

### **What if I want to know more?**

For further information regarding areas the present study is concerned with, please see the following papers:

Aubeeluck, A., Buchanan, H. & Stupple, E. N. (2012). 'All the burden on all the carers': exploring quality of life with family caregivers of Huntington's disease patients. *Quality of Life Research*, 21, 1425-1435.

Cairns, V., & Murray, C. (2015). How do the features of Mindfulness-Based Cognitive Therapy contribute to positive therapeutic change? A meta-synthesis of qualitative studies. *Behavioural and Cognitive Psychotherapy*, 43 (3), 342-359. doi: 10.1017/S1352465813000945.

Paulsen, J.S., Nehl, C., Hoth, K.F., Kanz, J.E., Benjamin, M., Conybeare, R., ... & Turner, B. (2005). Depression and stages of Huntington's disease. *Journal of Neuropsychiatry and Clinical Neurosciences*, 17 (4), 496-502.

Williams, J. K., Skirton, H., Paulsen, J. S., Tripp-Reimer, T., Jarmon, L., Mcgonigal Kenney, M., Birrer, E., Hennig, B. L. & Honeyford, J. (2009). The emotional experiences of family carers in Huntington disease. *Journal of Advanced Nursing*, 65, 789-798.

If you have concerns about your rights as a participant in this experiment or have any further questions, please contact Rachael Theed at [r.theed@lancaster.ac.uk](mailto:r.theed@lancaster.ac.uk)

Thank you again for your participation.