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

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**Abstract**

**Objectives:** This study sought to investigate how people who had tested positive for the Huntington's disease's (HD) gene mutation understood and experienced psychological distress and their expectations of psychological therapy.

**Design:** A qualitative methodology was adopted involving semi-structured interviews and interpretative phenomenological analysis (IPA).

**Method:** A total of nine participants (five women and four men) who had opted to engage in psychological therapy were recruited and interviewed prior to the start of this particular psychological therapeutic intervention. 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 in attributions of distress over time: *"in the past you'd just get on with it"*; Approaching therapy with an open mind, commitment and hope: *"a light at the end of the tunnel"*.

**Conclusion:** Understandings of psychological distress in HD included biological and psychological explanations, with both often being accepted simultaneously by the same individual but with biomedical accounts generally dominating. Individual experience seemed to reflect a dynamic process whereby people's understanding and experience of their distress 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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### **Practitioner points**

- People with the Huntington gene mutation have largely biomedical understandings of their psychological distress.
- This largely biomedical understanding does not, however, preclude them for being interested in the potential gains resulting from psychological therapy.
- The mechanisms of psychological therapy should be explained in detail before therapy, and explored along with current attributions of distress

### **Introduction**

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 mutated gene from an affected parent, people with HD have often seen their parents live with – and die from – 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). 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). Additionally, a diagnostic test is performed once a person presents with problems indicative of HD (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). Moreover, high levels of depression in HD, ranging from 33-69% (see Van Duijn, Kingma & Van der Mast, 2007 for review), have been reported, with depression most observed in HD when an individual's functional capacity and independence is affected (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. 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 & Predict-HD Investigators of the Huntington Study Group, 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 argued to be a core psychological feature of HD at the pre-symptomatic stage (Kloppel et al., 2010); it is hypothesised 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

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found in those at pre, early and advanced stages, indicating that people with HD may experience these for many years and, subsequently, may need support throughout the trajectory of the disease.

Many researchers take the view that psychological difficulties occur as a result of biological factors whereby neural mechanisms in the brain are affected by the gene mutation and these 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.. A key milestone for an individual is the result of the HD genetic test. A review of the impact of genetic testing found that distress was not consistently associated with test result (i.e. whether carrying the mutated gene or not) and that distress fluctuated over the years following the test result (with an initial increase in hopelessness with those who were gene mutation positive which later declined), although those with greater distress may be under-represented as they were more likely to be lost to follow up (Crozier, Robertson & Dale, 2015). Predictors of distress following a test result included prior distress in the person themselves or their family, the result itself and the expectation of the result (Crozier et al., 2015). Perhaps lack of a clear sustained negative effect of testing for all is unsurprising as qualitative research suggests a variety of emotional responses may be experienced following a positive test result. Such responses include shock, denial and anger, regret at taking the test, feeling that life is over, distress at anticipated life changes, worries about future relationships, fears of discrimination and loneliness but also relief from uncertainty, acceptance, closer relationships, a commitment to living in the moment and determination to make the best of life (Duncan, 2007; Gong, 2016; Hagberg, 2011; Schwartz, 2010).

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Thus, while studies generally indicate reaction to diagnosis (or genetic test result) is not the main cause of depression for people with HD (Craufurd & Snowden, 2014), the qualitative findings would suggest such knowledge could be important for some individuals (Hagberg, 2011; Schwartz, 2010), particularly if there is a history of psychological and social difficulties (e.g. Silver, 2003). Similarly, theories of anxiety in HD include pathological changes, but also the effects of possible cognitive overload along with familial and interpersonal factors (Dale & Van Duijn, 2015). In addition, the high rate of psychological difficulties in HD family members without the mutated gene may suggest that growing up at risk and/or in a family affected by HD also has a psychological impact ( e.g., Julien et al., 2007). Alongside social factors, 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).

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 (Aubeeluck & Wilson, 2008) and how this can impact on their other drug regimes. Consequently, psychological interventions may provide an alternative or additional way to reduce distress.

Currently, very little evidence exists on the acceptability and efficacy of psychological approaches for people with HD. For example, a single case study has highlighted the possible benefit of cognitive behavioural therapy CBT (Silver, 2003); also, a patient and carers education programme including sessions on developing coping strategies and psychosocial support (A'Campo, 2012) and 1- and 2-year multi-disciplinary intensive

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rehabilitation programmes have also shown some promise in alleviating distress or preventing deterioration in psychological functioning (Zinzi et al, 2007; Zinzi, Salmaso, Frontali & Jacopini, 2009; Piira et al., 2013; Piira et al., 2014). However no clinical trials or feasibility studies on psychological therapy specifically are currently published. 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). Clearly, then, as a provisional step, it could be useful to assess whether psychological interventions are seen as potentially beneficial by people with HD. Beliefs around the effectiveness of an intervention (or its mechanism of effectiveness) are important in determining its success and any psychological intervention which presupposes certain outcomes without more in-depth preparatory research runs the risk of poor uptake and high levels of attrition (Regan, Lambert & Kelly, 2013). For example, 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.

As a result, this study adopted a qualitative methodology in order to obtain detailed accounts of the understandings and experience of psychological difficulties and expectations of psychological therapy of individuals who are known to carry the HD gene mutation, but do not yet have a clinical diagnosis of HD. 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. Consequently, this study aimed to investigate individuals with the HD gene mutation's understanding of their psychological difficulties and their views

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of psychological therapy. Specifically, a potential pool of participants receptive to the concept of psychological therapy were chosen as potential participants given that these individuals would be more likely to take part in a psychological intervention and be more likely to participate in an exploratory study.

### **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). Before recruitment, ethical approval was gained from the UK National Research Ethics Service as well as approvals from the referring hospital.

#### **Participants and recruitment**

Participants were recruited from a specialist Huntington's disease clinic in the North of England and had to meet the following criteria: consultant-confirmed CAG expansion on the huntington gene and either pre-symptomatic or at an early stage. This study was introduced by the intervention coordinator and potential participants were given written information and time to decide whether to take part. In the current study all participants were required to understand and be able to speak English and be aged 18 or over.

Nine participants (recruited from 11 who were due to start a mindfulness intervention) agreed to take part, five of whom were female and four male. None were currently in receipt of a diagnosis. 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



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taking antidepressant medication and four participants had previous experience of psychological therapy. All participants - given pseudonyms in this study - had indicated their willingness to be involved in psychological therapy for low mood.

### **Data Collection**

Data were collected via individual interviews guided by a semi-structured schedule; question areas included: background probing on their current experiences of HD; their views of psychological therapy (what they considered this was and what had informed these views); views on different types of therapy; factors which influenced their decision to wish to engage in therapy; what their expectation would be of any change and the mechanisms of such change. Interviews were completed during October 2015. All interviews were conducted face to face by either the first or second author, and at community locations of the participants' choice, most usually their homes. Interviews lasted between 45 and 65 minutes ( $M = 54$  minutes). Audio recordings were transferred onto a computer as soon as possible and stored on the authors' university server via a secure link, password protected and then deleted from the recording device. Once the data had been transcribed, checked and analysed, they were deleted.

### **Data analysis**

All interviews were transcribed verbatim and anonymised to protect participants' confidentiality. The data were analysed by the first author using IPA, following the stages outlined by Smith and Osborn (2003). For each participant their individual transcript was read and then re-read with comments relevant to the research question being noted and used to develop emerging themes. Following this, emerging themes were then clustered together based on their apparent similarities by copying the emerging themes into a table and giving

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each cluster a theme name. This was done individually for each participant. Once this was complete, super-ordinate themes were developed which best fit the majority of participants' experiences.

IPA was chosen due to its ability to offer both an individual and group level of analysis, to present participants' phenomenological understanding of their experiences and the meaning they ascribe to these and to allow the researchers to provide an additional level of interpretation (Smith & Osborn, 2003).

### Results

Analysis of the data resulted in the development of three themes. Each theme is described below.

#### **Attributing psychological distress to HD: *"you're blaming everything on that now"***

This theme describes how participants understood their experience of psychological distress. All participants appeared to attribute their psychological distress to HD; however this was from both a biological and psychological perspective.

Most participants described, more fully, a biological understanding of psychological difficulties in HD attributing difficulties such as low mood, anxiety and irritability to the disease process, 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).

Certainly, these explanations appeared to be strongly influenced by the discourse provided by participants' health care professionals, the majority of whom were medically trained. As Alice explained "they [professionals] 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

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depressed". It also emerged that this biological explanation of psychological distress, to which participants were more frequently exposed, was also acceptable to participants. 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...I do tend to think it's a biological thing. You know I've got a really good life you know, I enjoy myself and I like who I am, so I can't understand otherwise why I would be down" (Sharon). It seemed that this participant could only understand her mood from a biological perspective as, to her, nothing else in her life could be responsible for it. Consequently, in her opinion attempts to control or modify her mood were limited.

Although not rejecting a biological understanding, some participants were more ambivalent about the cause of low mood if this ambivalence was reflected in comments made by their health care professionals. Lyndsey's experience was that "they [medical professionals] often say they don't know if depression's linked to HD and that they don't know either way". This then gave her permission to have a more nuanced understanding: "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]". For many participants it appeared that both psychological and biological explanations of distress existed in parallel.

Many participants seemed to experience their HD premanifest status as directly influencing 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, the nature of the range of difficulties which participants might go on to experience was emotionally very unsettling; as Lyndsey said: "Sometimes I depress myself because sometimes maybe I do think too far ahead...HD just affects so many aspects and that does scare me."

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"

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(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 their gene status seemed to result in the replacement of one type of ambiguity – about their gene status – with another one: how and when they would be affected by the condition. This ambiguity was often experienced as leading 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 ... you'll never know definitely ... it's not a set path which is really hard" (Alice).

Additionally, the understanding that the experience of physical and psychological difficulties was perceived to be inevitable (due to its genetic transmission) also resulted in distress:

"Sometimes I depress myself because sometimes maybe I do think too far ahead...HD just affects so many aspects and that does scare me" (Lyndsey)

Additionally, many participants talked about their experience of psychological distress, in particular anxiety, in the context of worrying about the genetic transmission of HD: "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. In order to manage this cause of distress, avoidance was often used due to an inability to control this possibility.

All participants described their experience of HD as a loss of control, which then resulted 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 a sense that participants 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

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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 they had limited treatment options, increasing their feelings of helplessness.

### **Changes in attributions of distress across time: "*in the past you'd just get on with it*"**

The receipt of the positive test result had radically changed participants' perception of the significance and cause of their psychological difficulties. Participants were now highly sensitive to any instance of psychological distress, "whereas in the past you'd just get on with it" (Chris). Moreover, when people experienced difficulties following the positive test result, it seemed that they were increasingly likely to attribute these to the biological 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".

Another specific change in attributional process regarded the origin of emotional responses and whether these were part of participants' personality or the effect of the HD gene mutation. 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 lack of clarity was further highlighted by Sue who discussed her experience of irritability and anger as "part of the condition" but then 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 their gene status had provoked a re-assessment of 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

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his personality, however with the potential for HD to accentuate this: "I don't think the HD's brought them [emotional difficulties] 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 most participants' understanding of their experience and distress, not only regarding their present and future but also their past. Only one participant continued to see his experience of psychological distress prior to and following confirmation of his gene status as separate. On discussing experiencing a period of depression prior to knowing about HD, Simon commented "I don't attribute any of that to HD related stuff, that was really to do with work pressures".

### **Approaching therapy with an open mind, commitment and hope: *"a light at the end of the tunnel"***

Most participants described seeing the opportunity to engage in psychological therapy as positive, even for those who had had previously unhelpful experiences of therapy ("I did go but I didn't really get it", Sue). Moreover, most participants were not sure what to expect but hoped 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 their gene status 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 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

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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. Their preference seemed to originate from their belief that medication involved introducing external agents 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.

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.

However, despite the hope people had for psychological therapy to help them with the psychological difficulties they experienced, due to their biological understanding of psychological difficulties, 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. This was further reflected in comments from some participants who expressed an understanding and expectation that psychological therapy 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

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worse...there's probably a limit to what it can do" (Chris). Indeed, it seemed that participants 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 these could 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).

An understanding that engaging in psychological therapy required a certain mind set in order for the therapy to be beneficial was also evident, 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 important due to the 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 et cetera.  
(Sharon)

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 engaging actively 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



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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.

### **Discussion**

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 variable, 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, i.e., a response to the challenges of living with HD. However, no other psychological explanations, for example, around the difficulties of living in a family where a parent has HD, were proffered. Moreover, the more dominant understanding, running alongside a psychological account, attributed psychological difficulties to the biological process of HD and, subsequently, were extremely likely to occur.

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

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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 of psychological distress seemed to reflect a dynamic process for many participants as opposed to being static. This was particularly the case following the genetic test when attributions for distress became more dominated by HD. The self-regulation model of chronic illness (Leventhal, Meyer and Nernez, 1980) proposes that people, based on their experience of their illness, develop their own illness beliefs. These include perceived symptoms and perceptions of cause, control and consequences as well as the likely temporal course of the illness, to help them make sense of their illness and subsequently cope with and adapt to it. Indeed it has been suggested that people's beliefs about their illness are often influenced, unsurprisingly, by the information to which they are most exposed and as such these beliefs change (Leventhal, Leventhal & Cameron, 2001).

On learning that they had the mutated gene and that depression was a symptom of HD, it is unsurprising that for several participants the dominant attributions for distress changed. Furthermore, as HD is incurable, with limited options for symptom management (Walker, 2007), any low mood could also be similarly pessimistically appraised (i.e. their control beliefs for depression mirrored those for HD). Furthermore, these beliefs could influence attempts to manage any distress. As one participant observed, in the past he just "got on with it", whereas if now the cause was biological (with no cure), beliefs in ability to manage distress are reduced (except perhaps by medication). Nonetheless, during the interviews, a more subtle understanding emerged as participants acknowledged roles for other psychological factors including a reaction to the diagnosis, living with uncertainty, concerns about heritability of the condition and part of their "personality". Interestingly participants did not suggest their backgrounds, including growing up in an HD family, might be a possible causative factor (Julien et al. 2007; Silver, 2003).

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In a systematic review examining the psychological impact of predictive testing, an initial increase in feelings of hopelessness was found, although this decreased in later years (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 affect 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.

In addition, the current findings support those of Arran et al. (2013) who found that people with the HD gene mutation 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, thus taking a proactive approach with regards to their wellbeing, appeared to enable participants to feel they were regaining some of the control they had lost. Thus increasing a person's perception of control over some aspects of 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 the gene mutation will hold positive control beliefs. Consequently, it may be more effective to work with people with the condition to accept and learn to live with the reduced control and the ambiguity HD brings. Certainly, living with the unpredictable and uncontrollable nature of this condition, 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).

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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 mutation easier than the uncertainty of HD, particularly so for younger people (Gong et al., 2016; Duncan et al., 2007). While this was true for the majority of participants, this then resulted in a different uncertainty that people had to manage (see also Hagberg et al., 2011). 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 (Duisterof, Trijsburg, Niermeijer, Roos & Tibben, 2001). Subsequently, there were wider implications of having the HD gene mutation than just those of the individual. Indeed, anxieties were discussed as a result of the potential to have passed on the gene, similarly to Hagberg et al.'s (2011) participants. Furthermore most people, given its genetic transmission, will have seen a family member 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 may be, anxiety levels may be expectedly high.

Previous studies have shown patient outcome expectations to be important in engagement and completion of psychological therapy programmes (e.g., Snippe et al., 2015), with one study's authors commenting: "outcome expectations reflect patients' prognostic beliefs about the consequences of engaging in treatment" (Constantino, Arnkoff, Glass, Ametrano & Smith, 2011, p.184). In addition, context has been suggested to be a potential

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influence on an individual's expectations insofar as if a person has prior experience of a psychological therapy then their expectations of future therapy may be influenced by their previous experience (Constantino et al., 2011). 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 its utility. Indeed, despite some participants having a negative experience of previous therapy, this did not seem to influence their hopeful expectations of future interventions.

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 individuals have a tendency to seek consistency regarding their cognitions (i.e. beliefs). When there is not consistency, dissonance occurs. 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 mitigated by hope. Even though dissonance can occur between a person's beliefs and their actions (i.e. holding a biological understanding and accessing psychological therapy), this is tolerable when the potential benefit of the dissonant action has the potential to outweigh this conflict.

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

The current research highlights a number of important clinical implications for developing therapies for people with the HD gene mutation. Firstly, it has been shown that this group are open to a psychological understanding of distress and – even while also holding biological beliefs about causation – would welcome subsequent psychological

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approaches. Consequently, the provision of, and access to, psychological therapy services for people with pre-manifest and manifest HD should be actively pursued. Indeed, other findings (Tabrizi et al., 2012; Eidelberg & Surmeier, 2011) support the argument for non-pharmacological approaches such as CBT for support with 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 and either as a result or a reason for this – or both – there are currently no clinical trials or even feasibility studies assessing the effectiveness of specific psychological approaches in people affected by Huntington's. However, here there is an indication that psychological approaches may be acceptable to people with HD with the potential to improve well-being.

Furthermore, these results indicate the importance of instilling hope for positive psychological outcomes, especially given the biological explanations so often used to explain psychological difficulties in HD, and for referring clinicians to take time to explain the mechanisms by which psychological therapies can exert a therapeutic effect. This explanation needs to be couched within models which suggest that evidence suggests that even biologically mediated mood changes can often be alleviated by psychological therapies. Finally, it is important to stress that psychological therapies differ and certainly 'one size does not fit all'. This means that even participants for whom therapy has not been effective previously should be aware that different approaches exist and, just as with physical therapies or medication regimes, these can be effective for different individuals.

A number of limitations exist in this study. 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

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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 study has demonstrated that participants accepted both a biological and psychological understanding of psychological distress, however with a biological perspective seeming to dominate. Furthermore, participants' attributions were 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 by the hope that this could provide an alternative or additional approach to medication that could support people with HD to feel more in control of their experience.

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