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Couples' illness representation and coping procedures in prodromal Huntington disease

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COUPLES' ILLNESS REPRESENTATIONS AND COPING PROCEDURES IN PRODROMAL HUNTINGTON DISEASE

by

Nancy Ruth Downing

An Abstract

Of a thesis submitted in partial fulfillment
of the requirements for the Doctor of
Philosophy degree in Nursing
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Thesis Supervisor: Professor Janet K. Williams

ABSTRACT

Huntington disease (HD) is a degenerative neurological disease that leads to severe impairment in cognitive, behavioral, and motor function and premature death. Persons who test positive for the HD gene expansion know they will develop the disease, typically in mid life. Research indicates changes are detectable several years before onset of distinctive motor symptoms. The period of time between a positive test result and diagnosis has thus been called prodromal HD (prHD). Little is known whether persons with prHD or their companions notice changes, or how they cope with them.

The purpose of this thesis was to use the Common Sense Model to explore and describe illness representations in persons with prHD and their companions in three papers. The first paper was a preliminary analysis of interview data from eight persons with prHD and seven companions. Results indicated participants noticed and made attributions for changes in work function but were unsure whether some changes were related to HD. Results were preliminary because participants were not asked to make attributions and the sample size was small.

In the next two papers, 23 couples were interviewed. The purpose of the second paper was to explore illness representations in persons with prHD and their companions and evaluate the usefulness of the CSM in anticipated illness. Results supported preliminary findings: Participants noticed changes and made attributions; again, most did not attribute them to HD. Participants also used and evaluated coping strategies. Other elements of the CSM were partially supported.

The third paper used mixed methods to explore coping in persons with prHD and companions. Participants were asked open-ended questions about how they coped with changes and were verbally administered the Brief COPE scale. Participants used active coping, acceptance, planning, and social support and rarely used denial or substance abuse. Persons with prHD used more coping strategies than companions. Three major

themes from the qualitative included: trying to fix it, can't fix it, and not broken yet. Interviews revealed some coping strategies the Brief COPE did not measure. Findings from these papers may inform interventions to help persons with prHD and companions cope with changes.

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CERTIFICATE OF APPROVAL

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PH.D. THESIS

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To the brave and generous people from families affected by Huntington disease who are willing to participate in research

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INTRODUCTION

Background

Problem Statement

Huntington disease (HD) is a genetic disorder which leads to significant impairment in cognitive, behavioral, and motor function and premature death (Walker, 2007). Usually HD does not cause significant impairment until midlife with average age of onset of distinctive motor signs between 35 and 55 (Quarrell, 2008). Diagnosis of HD is made based on presence of definitive motor signs: chorea, dystonia, and impaired voluntary movements (Hogarth, 2003). However, at-risk individuals can undergo predictive testing prior to diagnosis.

A growing body of research indicates that changes in cognitive and motor function are detectable in persons with the HD gene expansion at least 15 years prior to diagnosis (Paulsen, 2010). The period prior to diagnosis has often been referred to in the literature as "presymptomatic" (Witjes-Ane, et al., 2007). However, recent evidence suggests a more appropriate term to acknowledge early changes is "prodromal HD" (Paulsen, 2010, p. 85). The term prodromal HD (prHD) is thus used throughout this dissertation, although an earlier term, "pre-HD" was used in Chapter 2.

Despite the increased knowledge of changes prior to traditional diagnosis of HD, little is known regarding whether persons with prHD or their companions notice changes, or if they do notice changes, whether they attribute them to HD. Family members state they don't know whether changes in persons with prHD are related to HD and express a desire for more information from healthcare providers regarding what to expect (Williams et al., 2007). According to Leventhal and colleagues' Common Sense Model of Illness Representation (CSM), the way people make sense of somatic changes is important because it influences how they cope with them (H. Leventhal, Meyer, &

Nerenz, 1980). If people select coping procedures that are not effective, this has a negative impact on their wellbeing. Thus coping modulates wellbeing. Therefore, it is important to explore how persons with prHD and their companions make sense of changes and how they cope with them.

From a developmental perspective, changes may begin to interfere with functioning at a time of life when people are engaged in generative activities, including careers and raising families. Changes that interfere with these activities have the potential to be very distressing both to persons with prHD and their companions. Thus, it is important to explore whether they notice changes and what they do to cope with them. These findings could be useful in developing interventions to help people cope better during the prodromal period.

Purpose and Aims

The purpose of this study was to explore illness representations and coping procedures of persons with prHD and their companions. The following specific aims were addressed in three papers:

1. Describe the attributions that persons with prHD and their companions made for functional changes in prHD and the active processes used to make attributions;
2. Explore illness representations in persons with prHD and companions using CSM and evaluate the appropriateness of the CSM in anticipated illness using prHD as a model;
3. Describe the coping strategies used by persons with prHD and their companions to manage changes.

Huntington Disease Overview

George Huntington provided the first detailed description of HD in his 1872 paper "On Chorea." Huntington was a physician on Long Island, in a community with several HD-affected families. While the term "chorea" (Greek for "dance") had been used

previously to describe any number of diseases characterized by involuntary movements, Huntington provided a detailed description of "hereditary chorea" (p. 111). He noted that it did not "skip a generation" (p. 112) and if one generation was not afflicted, the disease would no longer manifest in that lineage. Thus, while having only a rudimentary understanding of heredity, Huntington identified what is now known as a dominant inheritance pattern.

For many years the disease was referred to as "Huntington's chorea" in reference to George Huntington. Today researchers and clinicians recognize that chorea is only one type of movement disorder in HD, and the disease is also characterized by behavioral and cognitive changes. For these reasons, the preferred term is Huntington disease (Quarrell, 2008). Chorea may be subtle initially and resemble fidgeting. Gradually, however, the involuntary movements become larger and more pronounced; problems with balance occur (Quarrell, 2008). Other motor symptoms that increase with disease progression are dystonia and impaired voluntary movements, including bradykinesia (Quarrell, 2008). Dystonia refers to holding the limbs in unusual positions. It becomes more difficult for people with HD to control voluntary movements as the disease progresses and movements become slowed (bradykinesia). Eventually, people with HD can become rigid and have difficulty swallowing. Swallowing difficulties may contribute to aspiration pneumonia, a common cause of death in HD (Dubinsky, 2005).

Huntington disease is autosomal dominant, which means that offspring of affected individuals have a 50% chance of developing the disease (OMIM, 2010b).The disease involves a trinucleotide (CAG) expansion of the huntingtin gene (HTT) on chromosome 4 (OMIM, 2010b). Age of diagnosis is associated with the length of the gene expansion (Langbehn et al., 2004), although wide individual variations exist. The average lifespan following diagnosis is 17-20 years (R. H. Myers, 2004). The prevalence of HD in North America is approximately 1 in 10,000 (Walker & Raymond, 2004), although it may be underestimated due to stigma (Wexler, 2010).

Since the gene that codes for HD was discovered in 1993 (The Huntington's Disease Collaborative Research Group, 1993), at-risk individuals can undergo predictive genetic testing prior to diagnosis (R. H. Myers, 2004). Persons who will not develop HD have two normal HTT alleles with ≤26 CAG repeats on each allele (Human Genetics Society of Australasia, 2001). People with 27-35 CAG repeats on at least one allele will not develop HD, but it is possible their offspring will because the repeat is unstable in some people; an unstable repeat can expand in subsequent generations, usually when transmitted by a male (R. H. Myers, 2004). People with >35 CAG repeats will develop HD if they live long enough. The CAG repeat on the HTT gene codes for glutamine. Excessive glutamine creates an abnormality in the associated huntingtin protein which is associated with premature neuronal cell death (Walker, 2007). The mechanisms behind these changes are not yet fully understood; however, brain imaging in persons with prHD indicate damage in several areas of the brain, including the basal ganglia (Aylward, 2007; Beglinger et al., 2005), white matter (Paulsen et al., 2010; Stoffers et al., 2010), and the cortex (Nopoulos et al., 2010).

Behavioral, cognitive and motor symptoms become progressively worse and in the later stages include dementia, rigidity, and difficulty swallowing (Quarrell, 2008). The most common causes of death in HD include pneumonia (often related to aspiration due to impaired swallowing) and cardiovascular disease; other causes include cachexia, suicide, and accidents (Sorensen & Fenger, 1992). Currently there is no cure for HD; however, treatments for symptoms exist (Adam & Jankovic, 2008; Mason & Barker, 2009).

Behavioral and psychiatric changes in diagnosed HD include depression (Paulsen et al., 2005), anxiety (Marshall et al., 2007), apathy (Kirkwood, Su , Conneally, & Foroud, 2001; van Duijn, Reedeker, Giltay, Roos, & van der Mast, 2010), obsessive-compulsive symptoms (Beglinger et al., 2008), irritability (Kingma, van Duijn, Timman, van der Mast, & Roos, 2008), and aggression (Cummings, 1995). Cognitive changes

include slowed thinking, impaired memory and executive function (Robins Wahlin, Lundin, & Dear, 2007), and diminished insight (Hoth et al., 2007). While definitive predictive testing for the HD gene expansion is now possible, uptake prior to motor symptom onset is estimated to be between 3-24% of at-risk individuals (Tibben, 2007).

Persons who have undergone predictive testing for the HD gene expansion cited various reasons for their choice: relief from uncertainty (Richards, 2004), knowledge and understanding (Williams, Erwin, Juhl et al., 2010a), future planning, reproductive decisions, and to inform children who may be at risk (Decruyenaere et al., 2003; Meiser & Dunn, 2001; Williams, Erwin, Juhl et al., 2010a), and to obtain social support (Williams, Erwin, Juhl et al., 2010a). Persons who chose *not* to be tested also cited various reasons: a desire to conceal their own or family members' risk of HD, to preserve hope and optimism (Quaid et al., 2008; Williams, Erwin, Juhl et al., 2010a), fear they will not be able to cope well with the information (Codori, Hanson, & Brandt, 1994), and fear of genetic discrimination (Erwin et al., 2010).

Prodromal Huntington Disease

The ability to determine who will develop HD prior to diagnosis has created a unique opportunity to study the disease before the onset of distinctive motor symptoms. The search for clinical markers in prHD is underway in order to provide benchmarks for evaluating future treatments and attempts to cure HD (Paulsen, 2010). In the meantime, this research is generating a more accurate picture of the slow progressive course of HD.

In prHD, symptoms may be subtle and ambiguous initially and slowly progress until they become more distinctive. Some of the earliest noticeable changes may be psychiatric and behavioral changes (Duff et al., 2007). Family members report noticeable changes in the behavior of persons with prHD, especially irritability (Williams et al., 2007). Persons with prHD have demonstrated higher scores on measures of irritability, obsessive-compulsive symptoms, anxiety, and depression than people at-risk who tested

negative for the HD gene expansion (Beglinger et al., 2008; Berrios et al., 2002; Duff et al., 2007; Marshall et al., 2007). Some family members notice changes in the behavior, thinking, relationships and physical health of persons with prHD (Williams et al., 2007). They notice depression, moodiness, and decreased social activity and relationship quality; they also notice memory loss, poor judgment, motor and balance problems, and sleep changes.

Researchers have found impaired memory and executive function in persons with prHD compared with subjects with CAG repeats <36 using standardized cognitive batteries (Johnson et al., 2007; Rowe et al., 2010; Solomon et al., 2008). Subtle motor changes, including lower scores on tests involving finger tapping, tandem gait, saccade initiation, and mild chorea have also been detected (Biglan et al., 2009). Persons with prHD and their families may not be aware of these changes.

Changes in brain structure of persons with prHD have been observed using single photon emission computed tomography, magnetic resonance imaging (MRI) and functional MRI (Aylward, 2007; Harris et al., 1999; Reading et al., 2005; Zimbelman et al., 2007). These studies indicate changes in the basal ganglia, including loss of volume indicative of atrophy, and consistent with neuronal cell death. The basal ganglia are responsible for the coordination of voluntary movements (National Institutes of Health, 2007). Furthermore, basal ganglia volume has been shown to decrease over time as participants with prHD approach HD diagnosis (Zimbelman et al., 2007), and has been associated with decline in cognitive test abilities (Beglinger et al., 2005). Other areas of the brain have also been shown to be affected, including white matter (Paulsen et al., 2010) and the cerebral cortex (Nopoulos et al., 2010) which may account for some of the early changes seen in prHD.

The Common Sense Model of Illness Representation

The CSM provided a framework to explore how persons with prHD and companions made sense of changes in prHD and selected coping procedures to address them. According to the CSM, when people perceive somatic changes or receive health information, they try to make sense of these by developing an illness representation (H. Leventhal et al., 1980). Illness representations are defined as "individuals' common-sense definitions of health threats" (H. Leventhal, Leventhal, & Contrada, 1998), p. 719). Further, illness representations influence how people cope with perceived changes. The "common-sensical" connection between illness representations and the selection of coping procedures prompted the nomenclature "Common Sense Model" (H. Leventhal et al., 1998), p. 722).

The CSM is also referred to as the self-regulatory model because individuals use their perceptions of their inner states and external environment to guide their illness representations; they select coping procedures based on these perceptions, evaluate them and adjust their illness representations and coping procedures in an iterative process (H. Leventhal, Nerenz, & Steele, 1984). For example, if a coping procedure is ineffective, a person adjusts the illness representation or selects a different coping procedure. This process continues until the person evaluates the coping procedure as effective.

Formation of illness representations is an active process that involves cognitive and emotional ("parallel") processing, recursive stages ("representation" and "appraisal") and concrete and abstract thinking ("hierarchical processing") (H. Leventhal et al., 1984, pp. 219-220). Illness representations are composed of five attributes: identity, cause, timeline, consequences, and cure/controllability (Scharloo & Kaptein, 1998). Identity refers to labels and symptoms; cause refers to what underlies changes; consequences refer to how serious the illness is; cure/controllability refers to how much control one has over an illness, including whether it can be treated or cured. These attributes are interrelated; perception of one attribute impacts the perception of others. Attention to physical cues,

individual beliefs, and social comparisons all contribute to the active process of forming an illness representation (H. Leventhal et al., 1980; H. Leventhal et al., 1984). Illness representations are also influenced by past experiences with similar symptoms or illnesses, by health care providers, and by the broader culture (H. Leventhal et al., 1984).

Another component of the CSM is the "symmetry rule." This rule states that people seek labels for perceived symptoms, and when given a label, people seek symptoms to match (H. Leventhal, Benyamini, Brownlee, & Diefenbach, 1998, p. 25). The researchers demonstrated this phenomenon in a study whereby college students with normal blood pressure were told they had hypertension; the students began to describe symptoms of hypertension, including palpitations, dizziness, tension, and headaches, while their blood pressure did not change (Baumann, Zimmerman, & Leventhal, 1989). More recently it has been demonstrated in a study of Gulf War veterans (Brewer, Hallman, & Kipen, 2008). The researchers found correlations between war exposures and medical symptoms with evidence of both recall of war exposures preceding symptom report and vice versa. The symmetry rule suggests that persons who have tested positive for the HD gene expansion may be more likely to attribute perceived changes to HD because they know they are going to develop the disease.

It may be possible for healthcare providers to alter inaccurate illness perceptions that interfere with effective coping. For example, healthcare providers have empowered patients by helping them accept aspects of their illnesses that cannot be controlled (Aujoulat, Marcolongo, Bonadiman, & Deccache, 2008). Patients were more satisfied when physicians discussed illness representations with them (Frostholm et al., 2005). Patients provided more detail about their health concerns when physicians talked to them about their illness representations, resulting in improved communication regarding how to manage concerns (de Ridder, Theunissen, & van Dulmen, 2007). Effective interventions to improve coping by altering illness representations have been reported (Broadbent, Ellis, Thomas, Gamble, & Petrie, 2009b; Keogh et al., 2007).

Leventhal and colleagues did not develop a quantitative measure of illness representation. However, others have created questionnaires based on their work, including the Implicit Models of Illness Questionnaire (Turk, Rudy, & Salovey, 1986), the Illness Perception Questionnaire (Weinman, Petrie, Moss-Morris, & Horne, 1996) and Revised Illness Perception Questionnaire (Moss-Morris et al., 2002). These tools were designed to examine the five dimensions of the CSM. They are less useful in prHD because changes are often subtle and ambiguous. Furthermore, they don't allow researchers to capture the active processes involved in forming illness representations. Therefore, this study used qualitative methods to explore the process of forming illness representations in prHD. Prior researchers have explored illness representations using the CSM in persons with diagnosed HD (Helder, Kaptein, Van Kempen, Weinman, Van Houwelingen et al., 2002a; Helder, Kaptein, Van Kempen, Weinman, Van Houwelingen et al., 2002b; Kaptein et al., 2007). However, little is known about how people with prHD and their companions make sense of and cope with these changes. This study addressed these gaps.

In prHD, persons likely do not consider themselves ill at all. Thus, the use of an illness representation model in this context may seem presumptuous. However, the authors of the CSM believe their model is also applicable to *anticipated illness* (Brownlee, Leventhal, & Leventhal, 2000, p. 386), although they concede this has not been adequately tested. This study adds to the understanding of the CSM in the context of anticipated illness.

Coping

In the CSM, illness representations are important because they influence the coping procedures people select (H. Leventhal et al., 1984). The authors defined coping procedures as "the cognitive and behavioral actions we take (or do not take) to enhance health and to prevent, treat…and rehabilitate from illness" (H. Leventhal et al., 1998, p.

722). After using coping procedures, people evaluate them for their effectiveness. If they are not effective, people select new coping procedures and/or adjust their illness representation to facilitate more effective coping procedures. Thus, coping modulates outcomes.

Coping procedures include whether to seek medical care for noticed somatic changes (Cameron, Leventhal, & Leventhal, 1995). When people's illness representations are inaccurate, this may result in less effective coping procedures, including a delay in seeking medical care. For example, some people who have had transient ischemic attacks delayed seeking medical care because they failed to recognize symptoms were indicative of a serious health problem (Sprigg, Machili, Otter, Wilson, & Robinson, 2009). Illnesses often begin with subtle and ambiguous symptoms, resulting in a long threat appraisal period in which people are likely to make more benign attributions (H. Leventhal et al., 1998). Thus, people may wait until symptoms are more severe and distinctive before they seek medical care (Cameron et al., 1995). People may delay medical treatment until other coping procedures, such as the use of over-the-counter medications, are no longer effective (Reed, Rayens, Winter, & Zhang, 2008).

In the CSM, denial is assumed to interfere with the selection of appropriate coping procedures. However, others have argued denial can be protective for people with severe illnesses for which there is little hope for cure and little control (Lazarus, 1999). This may be relevant in HD, for which there is no cure, treatment is limited, and persons with prHD face an inevitable decline in health that results in profound disability and premature death (Tibben, 2007).

Coping and Quality of Life

In the CSM, if people select effective coping procedures, this leads to positive outcomes (H. Leventhal & Colman, 1997). Implicit in this theory is the concept of quality of life, which is enhanced when coping procedures are effective. Lazarus and Folkman

(1984) explicitly tie coping to quality of life: "Simply put, the quality of life and what we usually mean by mental and physical health are tied up with the ways people evaluate and cope with the stresses of living" (p. 181). In their assessment, physical functioning in work and social life, life satisfaction, and physical health are the expected outcomes of effective coping. Thus, functioning is an important component of quality of life. Wilson and Cleary (1995) developed a model of health-related quality of life that also includes the importance of functioning. They defined health-related quality of life as "aspects of quality of life that relate specifically to a person's health" (p. 60), involving physical, social, and role functioning, mental health, and general health perceptions. This definition is thus appropriate for use in this dissertation because it relates health-related quality of life to functioning and acknowledges the role of health perceptions.

Decline in functional capacity has been demonstrated to significantly decrease health-related quality of life for persons with diagnosed HD (Helder, Kaptein, van Kempen, van Houwelingen, & Roos, 2001). For these persons, loss of function may be more closely related to decreased quality of life than motor symptoms or cognitive function alone (Ho, Gilbert, Mason, Goodman, & Barker, 2009). One group of researchers has explored the relationships between illness representation, coping, and quality of life in diagnosed HD for both participants with HD as well as their spouses (Helder, Kaptein, Van Kempen, Weinman, Van Houwelingen et al., 2002a; Helder, Kaptein, Van Kempen, Weinman, Van Houwelingen et al., 2002b; Kaptein et al., 2007). They found that illness perceptions and coping procedures significantly predicted quality of life for both participants with HD and their spouses. In their conceptualization, coping played a mediating role between illness representations and quality of life.

Coping Procedures

While Leventhal and colleagues discussed the importance of illness representations in selecting coping procedures, they did not elaborate on specific coping

procedures. The quantitative instruments designed to measure illness representations do not include coping procedures. However, there is an extensive body of literature by other authors related to coping. Some of the most influential work on coping is by Lazarus and Folkman. They defined coping as "constantly changing cognitive and behavioral efforts to manage specific external and/or internal demands that are appraised as taxing or exceeding the resources of the person" (Lazarus & Folkman, 1984, p. 141).

In Lazarus' model, coping is a process that consists of three stages: forming a primary appraisal of threat; forming a secondary appraisal of potential responses to threat; and responding to threat (Lazarus, 1999). Primary appraisal involves emotional elements, while secondary appraisal is largely a cognitive process. This process is followed by reappraisal of the original threat. In this way, the coping process parallels the CSM in that it is iterative and recursive.

In Lazarus and Folkman's model, coping is classified into two major categories—problem-focused and emotion-focused. Problem-focused coping involves cognitive processes such as obtaining information and forming action plans; emotion-focused coping involves addressing emotional responses to threats while not altering the threats themselves (Lazarus, 1999). Although these two types of coping have been characterized as dichotomous, and even hierarchical (with problem-focused coping being superior), the authors point out that there are situations in which it is more helpful to use emotion-focused coping. For example, using problem-focused coping in situations that cannot be changed can be harmful. Problem-focused and emotion-focused coping appear to be similar constructs to the cognitive and emotional processes in the CSM. However, while Lazarus and Folkman conceptualize them as distinct ways of coping, they are parallel processes in the CSM (H. Leventhal et al., 1984).

Coping Measures

Several tools have been developed to measure coping, including Folkman and Lazarus' (1980) Ways of Coping Questionnaire. The current version (Folkman & Lazarus, 1988) contains 66 items which are classified into problem-focused and emotion-focused coping. A limitation of the questionnaire is that factor analyses yield different results across samples (Schwartzer & Schwartzer, 1996). This makes comparison across samples impossible. Folkman and Moskowitz (2004) concede coping scales are contextual. Thus, narrative approaches should be used in addition to coping scales in order to identify context-specific ways of coping.

Another widely used coping measure is the COPE (Carver, Scheier, & Weintraub, 1989). The authors originally designed the 60-item scale based on problem-focused and emotion-focused coping and addressed what they believed were limitations in the Ways of Coping measure. They believed the Ways of Coping measure was missing domains, contained too much ambiguity, and was derived empirically instead of theoretically. They designed the COPE based on coping theory by Lazarus and Folkman as well as their own self-regulation theory and other coping research. The COPE includes 15 scales based on three categories of coping: problem-focused, emotion-focused, and "less useful" (p. 267). The scale was designed to to assess dispositional or situational coping depending on the context and aims of the study in which it is used. The COPE has also been criticized for yielding different factor structures across samples (Donoghue, 2007). Although originally conceived to measure problem-focused, emotion-focused, and maladaptive coping strategies, Carver (2007) now discourages people from using the measure to identify coping styles. Instead, he recommends people look at each scale separately and compare them with other variables in the particular sample of interest.

A major drawback of both the Ways of Coping and the COPE is length and thus potential for subject burden (Folkman & Moskowitz, 2004). Carver (1997) developed the Brief COPE, a shorter version of the COPE, to reduce subject burden and address other

limitations of the COPE. The Brief COPE consists of 14 scales containing (active coping, planning, positive reframing, acceptance, humor, religion, emotional support, instrumental support, self-distraction, denial, venting, substance use, behavioral disengagement, and self-blame). Each scale consists of two survey items. The Brief COPE has been used to evaluate coping in various illnesses, including perinatal depression (de Tychey et al., 2005), cystic fibrosis (Wong & Heriot, 2008), and mental illness (Meyer, 2001).

In a study using the CSM framework in persons with head and neck cancer, researchers used the Illness Perception Questionnaire and the Brief COPE together to illustrate the relationship between illness representations and coping (Llewellyn, McGurk, & Weinman, 2007). They found that patients who perceived more negative consequences of their illness were more likely to use planning coping strategies. Participants with strong emotional representations were more likely to use active coping and positive reframing. More negative consequences were also associated with denial, substance abuse, venting, and self-blame. This study illustrates how coping procedures and illness representations are related.

Several researchers point out the limitations of using quantitative measures to identify coping strategies. While quantitative measures are useful for systematically describing coping, narrative approaches may be more appropriate in situations in which little is known regarding how persons cope. Coping is a process; quantitative measures are not designed to capture active processes. Semi-structured interviews may yield more valuable information regarding the processes of coping and evaluation of coping, and coping in specific contexts (Coyne & Gottlieb, 1996). An exploration of coping in HD caregivers, for example, revealed that avoidance coping was common (Lowit & van Teijlingen, 2005). Many caregivers had ignored early symptoms of HD and delayed seeking medical care until symptoms were impossible to deny. Qualitative interviews also revealed that family caregivers of people with HD have also coped by seeking

comfort from other family members, anticipating the death of the HD-affected relative, and using prescription medications (Williams et al., 2009). Some of the coping procedures described by family members would not have been captured via pre-constructed questionnaires. Furthermore, coping questionnaires do not assess the active processes involved in the selection of coping procedures and how they are linked to representations of the illness itself. A qualitative approach is important to understand the links between these phenomena.

Coping and Huntington Disease

While illness representation, coping, and their impact on health-related quality of life have been explored in persons with diagnosed HD and their spouses (Helder, Kaptein, Van Kempen, Weinman, Van Houwelingen et al., 2002a; Helder, Kaptein, Van Kempen, Weinman, Van Houwelingen et al., 2002b; Kaptein et al., 2007; McCabe, Firth, & O'Connor, 2009), these concepts have not been studied in prHD. Several studies have explored distress following predictive HD testing, however. Two longitudinal studies indicated persons who tested positive for HD showed initial distress (hopelessness, intrusive thoughts, avoidance) immediately after testing, but distress decreased after approximately 24 months, and increased again 40 months after testing (Tibben, Timman, Bannink, & Duivenvoorden, 1997; Timman, Roos, Maat-Kievit, & Tibben, 2004). Companions showed similar patterns of distress following partners' HD testing (Tibben et al., 1997).

In cross-sectional studies that evaluated longer-term coping after HD testing, mild depression was present in 58% of persons who tested positive and 24% of persons who tested negative an average of 3.7 years after testing (Gargiulo et al., 2009). However, in another study, persons who tested positive did not meet criteria for depression an average of 3.75 years after testing (Lickleder, Wolff, & Barth, 2008).

While these studies were designed to evaluate the long-term impact of psychological adjustment to HD testing, they do not explore how persons with prHD and their companions cope with day-to-day functional changes prior to diagnosis. In light of growing evidence that indicates functional changes begin several years prior to HD diagnosis, it is important to understand how persons with prHD and their companions make sense of these changes. The attributions they give to changes influence the coping procedures they select. Coping procedures have the potential to impact quality of life.

Including Companions in Research

There are several reasons family members make valuable informants when assessing changes in persons with prHD. First of all, family members may be the first to notice changes in daily functioning of persons with prHD. In some cases, family members may feel as though they are the only ones who notice these changes (Williams et al., 2007). Furthermore, there is recent evidence that insight is impaired in persons with prHD (Duff, et al., 2010a). Another recent study demonstrated higher apathy scores in persons with prHD than in persons who tested negative for the HD gene expansion (van Duijn et al., 2010). Mild cognitive impairment may also impair insight (Davis, 2001), and has been found to be present in persons with prHD (Duff et al., 2010b). Due to these cognitive changes, companions can provide valuable collateral information on persons with prHD.

In addition, the lives of family members of persons with prHD are also heavily impacted by HD. They have reported similar rates of distress as those with prHD, and worry about their children inheriting the disease (Decruyenaere et al., 2005). After diagnosis, quality of life decreased for caregivers as well as for those with HD (Ready, Mathews, Leserman, & Paulsen, 2008). In addition, companions of persons with prHD also experienced disruption in their work and recreational lives as symptoms progressed (McCabe, Roberts, & Firth, 2008). Spouses of people with HD also felt distressed by the

loss of their marital relationship (Williams et al., 2009). The coping procedures selected by spouses of people with HD may impact quality of life for both members of a couple (Kaptein et al., 2007). For these reasons, it is important to include family members in research designed to explore functional changes in prHD and the coping procedures used to address them because family members may notice changes that persons with prHD do not notice and because they are also affected by these changes.

Significance

Recent evidence suggests it is possible to alter patients' and spouses' illness representations and facilitate improved healthcare outcomes in conditions with effective treatments such as diabetes (Lawson & Harvey, 2009) and heart disease (Broadbent, Ellis, Thomas, Gamble, & Petrie, 2009a; Broadbent, Ellis, Thomas, Gamble, & Petrie, 2009b; Petrie, Cameron, Ellis, Buick, & Weinman, 2002). It is unclear whether altering illness representations in persons with prHD and their companions will improve their wellbeing. However, because treatments for symptoms are available, it is possible that attributing functional changes to HD in the prodromal period may facilitate use of treatments to improve symptoms that interfere with functioning.

There is currently no cure for HD, though there are treatments available to address some symptoms (Adam & Jankovic, 2008; Mason & Barker, 2009; Mestre, Ferreira, Coelho, Rosa, & Sampaio, 2009). Some of these treatments may be beneficial in prHD. For example, while depression is higher in people with HD than in the general population (Paulsen et al., 2005) it has also been noted to be higher in persons with prHD than in individuals who tested negative for the gene expansion (Duff et al., 2007; Julien et al., 2007). The use of antidepressants is high in prHD (Rowe, et al., unpublished data). The search for novel treatments is ongoing and may depend on knowing at what point in disease progression that it is best to intervene (Georgiou-Karistianis et al., 2003).

Recent data also indicate persons with prHD reported impaired job function, ability to maintain finances, and decreased driving skills (Beglinger et al., 2010); these issues all can negatively impact quality of life. Theoretically, persons at risk for HD stand to benefit from the knowledge they will develop HD because they can seek treatment for symptoms when they are first manifested. However, this assumes persons will recognize subtle changes in functional abilities and attribute them to HD.

Current HD treatments that may improve quality of life for persons with prHD and their companions include medications to treat irritability (van Duijn, 2010), mood (Phillips, Shannon, & Barker, 2008), and ability to focus (Grimbergen & Roos, 2003). It may be important to target symptoms before associated changes in brain structure become too severe (Hannan, 2005). Brain imaging research indicates that over 50% of neuronal cell death has already occurred by the time of clinical diagnosis (Aylward et al., 2004). Therefore, the best point to intervene is likely to be years before diagnosis.

Some persons at risk for HD have experienced some relief from distress and uncertainty after being diagnosed with HD (Duncan et al., 2008). This indicates that for some persons with prHD, information regarding changes in prHD may help to relieve distress related to uncertainty regarding the meaning of changes. Persons with prHD and their companions may have to perceive changes in the prodromal period to be important (i.e. they significantly impact their quality of life) before they seek medical advice or accept treatment. If they do not perceive changes to be important, they may not be motivated to address them; they may be more likely to wait until symptoms become more severe. Earlier recognition of symptoms may provide more time for future planning, to make family and financial plans, and allow for more gradual rearrangement of family roles. The additional time to plan may help people cope better with these adjustments.

Despite the discovery that symptoms occur years prior to the onset of distinctive motor changes, there are few data indicating whether persons with prHD and their family members recognize changes or attribute them to HD. There is some evidence that persons

with prHD and their family members do notice changes in behavior and functional abilities in prHD (Williams et al., 2007); however, they express uncertainty whether changes are related to HD. In Chapter 2 of this dissertation, findings are presented that indicate persons with prHD and their companions noticed functional changes but most did not attribute them to HD. In order for persons with prHD to benefit from interventions to delay or prevent changes that lead to functional impairment prior to diagnosis, it may be necessary for them to recognize that they may be related to HD (Mason & Barker, 2009).

The current explored whether persons with prHD and their companions noticed changes, the attributions they made for changes, how they made attributions, and the coping procedures they selected to address changes. The results provide insight into whether persons with prHD and their companions attribute changes to HD and 2 how coping procedures differ according to whether they attribute changes to HD or to other factors. Earlier diagnosis of HD-related changes may provide opportunities to identify and treat symptoms that are subtle but which may impair functioning, including work and social functioning, and which may impact quality of life.

Ethical Considerations

Persons with prHD can be considered vulnerable populations due to the severity of HD and the potential to be continually asked to participate in research (National Institutes of Health, 1979). Because HD is a severe degenerative illness with no cure, people with the condition may have a therapeutic misconception that researchers can help them (Matutina, 2010). In addition, the questions in the current study had the potential to cause psychological harm by asking participants to consider whether changes may have been related to HD when they hadn't previously considered that possibility. The discovery of changes decades before diagnosis is relatively new. These findings suggest a paradigm shift may be underway regarding how HD will be diagnosed in the future.

There are no data at this time regarding whether persons with prHD and companions want to know about HD-related changes prior to diagnosis. This dissertation takes an initial step in addressing this issue by exploring what persons with prHD and their companions already believe about changes in prHD.

Methods

Design

The dissertation project as a whole used a mixed-methods design. Mixed methods design is defined as "the use of two (or more) research methods in a single study, when one (or more) of the methods is not complete in itself" (Morse & Niehaus, 2009). The major aims of the project were to explore and describe illness representation and coping in persons with prHD and their companions. The theoretical drive, therefore, was inductive, for which a qualitative approach is appropriate (Shepard, Orsi, Mahon, & Carroll, 2002). The core component of the project was qualitative and included a quantitative supplemental component for the purpose of systematically comparing coping strategies. Using notation created by Morse and Niehaus (2009, p. 25), a qualitative core component with a simultaneously-collected supplemental quantitative component is notated as: QUAL+quant.

Chapter 2 was a qualitative study that explored and described attributions for work function changes made by persons with prHD and their companions. These findings were derived from the volunteered responses of participants; attributions for work function changes were not solicited in the semi-structured interviews. Chapter 3 was a qualitative study that builds on the preliminary findings of Chapter 2 by specifically asking persons with prHD and their companions to describe functional changes and to what participants attribute them. Additionally, Chapter 3 explored the usefulness of the CSM in anticipated illness, using prHD as a model. Although this study used the

theoretical framework of the CSM, its aim was nevertheless inductive; there were no hypotheses regarding whether the CSM was appropriate in this population.

Chapter 4 was a mixed methods study designed to explore coping procedures in persons with prHD and their companions. This topic has received little exploration beyond measuring psychological responses to HD testing. Therefore, the theoretical drive was qualitative and the methods used were appropriate for a qualitative study. However, a short quantitative measure of coping was included in order to systematically explore and compare coping methods in persons with prHD and their companions and to characterize coping in this sample.

Sample

Purposeful criterion sampling, consistent with qualitative methodology (Patton, 1990; Sandelowski, 1995), was used to identify prospective participants. The samples for this dissertation project consisted of persons with prHD who were participants in the PREDICT-HD study, based at the University of Iowa. PREDICT-HD is a longitudinal study designed to track changes in prHD and identify biological and clinical markers of prHD that will be useful in future clinical trials (Paulsen, 2010). Participants in PREDICT-HD have independently undergone testing for the HD gene expansion as a prerequisite to participation. Persons with CAG repeat lengths of ≥36 are eligible to participate as persons with prHD, along with a companion who provides collateral data. PREDICT-HD consists of 32 sites in the US, Canada, UK, Europe, and Australia and currently includes 825 participants with prHD and their companions. Participants travel to a PREDICT-HD site annually to complete a battery of tests, including neurological testing, MRI and blood tests, as well as clinician-rated and self-rated tests of motor, cognitive, behavioral, and everyday functioning.

The sample in Chapter 2 was comprised of persons estimated to be far, midway, and near to HD diagnosis using an algorithm based on CAG length and current age

(Langbehn et al., 2004) and their companions. In Chapters 3 and 4, 23 persons with prHD who were estimated to be ≤15 years from HD diagnosis and their 23 spouses or significant others were invited to participate. The cutoff of ≤15 years from HD diagnosis was to account for the probability that functional changes beyond 15 years from diagnosis would be less noticeable if present (Paulsen, 2010).

Measures

Demographic data were collected at the beginning of the interview. These items included: age, gender, how long companions have known their partners with prHD, and whether persons with prHD have children. Interviews were conducted using a semi-structured interview guide developed from the literature and from prior interviews with persons with prHD and their companions. The interview script was reviewed and revised with input from experts in prHD and family research (J. K. Williams and J. S. Paulsen). The Brief COPE was administered following the semi-structured interview. See Appendix A and Appendix B for the interview guides and Appendix C for the Brief COPE measure.

Procedure

This project was approved by the University of Iowa IRB. Potential participants were identified in collaboration with a PREDICT-HD coordinator, and included persons who have previously indicated they are willing to participate in HD-related research in addition to PREDICT-HD. The researcher contacted potential participants by telephone or mail to describe the study and invite them to participate. Interested participants contacted by telephone subsequently received written information about the study and two copies of the consent documents each via mail. Mailed recruitment materials included a cover letter, two copies of the consent document per prospective participant, and a preferred contact form. When a participant returned a copy of the signed consent document in the provided stamped, addressed envelope, the researcher contacted them by

telephone to arrange a convenient interview time. Recruitment continued until all persons who wanted to participate were interviewed.

Data Management

Qualitative data for Chapters 3 and 4 were managed using strategies outlined by Knafl and Webster (1988) with the addition of the use of NVivo8 qualitative data management software (QSR International, 2000). Qualitative data management, according to Knafl and Webster is a reductionist activity in which interview transcripts are reduced to coded excerpts and organized according to descriptive themes. Descriptive coding categories were derived from the CSM framework and from results of the preliminary study and included: changes, attributions for changes, active processes of forming attributions, coping procedures, and evaluation of coping procedures.

Analysis

Descriptive analysis was used to analyze data for Chapter 3 (Sandelowski, 2000) and descriptive interpretive analysis was used to analyze data for Chapter 4 (Thorne, Kirkham, & MacDonald-Emes, 1997). Different analysis methods were used due to different aims: The aims of Chapter 3 were purely descriptive; the purpose of descriptive analysis is "to sensitize the reader to the viewpoint of a particular group," (Knafl & Howard, 1984, p. 20), which is important when presenting a topic about which little is known (Knafl & Webster, 1988). On the other hand, the aims of Chapter 4 were to move beyond pure description and create interpretive themes. Interpretive research acknowledges the active role of the researcher in creating meaning out of descriptive data (Lowenberg, 1993). The ultimate goal of descriptive interpretive analysis in nursing research is facilitate development of practical interventions to address health and illness concerns (Thorne et al., 1997).

Quantitative analyses included descriptive statistics: gender, mean age, mean number of years companion has known person with prHD, and percentage who have

children. Mean scores and standard deviations on the Brief COPE indicated how often participants used coping procedures; the purpose of this calculation was to explore how frequently persons with prHD and companions used coping procedures, an indication of whether current demands exceeded resources (Lazarus & Folkman, 1984). Means and standard deviations were calculated for each of the 14 coping scales to determine which coping procedures were used more frequently by persons with prHD and companions. Dependent *t*-tests were used to compare mean frequency of coping procedures between persons with prHD and their companions. Independent *t*-tests were used to test for differences between genders and between persons who attributed noticed changes to HD versus those who attributed changes to other things. Correlations between number of changes attributed to HD and frequency of coping procedures were also calculated to indicate whether more HD-related changes required more coping procedures.

Summary

Persons with prHD represent a unique population in which to explore illness representation. While they may not have been experiencing symptoms at the time they underwent HD genetic testing, persons who test positive for the HD gene expansion are faced with an illness threat they know will become a reality. A growing body of research indicates symptoms begin several years prior to HD diagnosis (Paulsen, 2010). While persons with prHD and their companions notice changes, it is unclear whether they attribute changes to HD. This is complicated by the possibility that some changes may in fact not be related to HD, but to other health conditions and life stressors. It may be important to sort out which subtle functional changes in prHD are related to HD and which are related to other factors in order to manage them appropriately. Researchers should also be aware of other stressors that contribute to changes in persons with prHD in order to control for potential confounders.

The exploration of these issues in this dissertation contributes to the understanding of how persons with prHD and their companions perceive functional changes in prHD, to what they attribute changes, how they make attributions, and how they cope with changes. These issues were explored via 3 aims: 1) Describe the attributions persons with prHD and their companions make for functional changes in prodromal HD and the active processes of making attributions; 2) evaluate the appropriateness of the CSM in anticipated illness using prHD as a model; 3) describe the coping procedure selected by persons with prHD and their companions to manage perceived changes in prHD.

These aims are presented in the form of three papers as Chapters 2, 3, and 4. Chapter 2 presents a data analysis from a larger mixed methods study designed to create a measure of work function changes in prodromal HD. Chapters 3 expands on the preliminary findings in Chapter 2. Chapter 4 moves beyond noticing changes and attributions to explore coping in prHD. Chapter 5 includes discussion and conclusions, limitations of the dissertation study, and clinical and research implications.

CHAPTER 2

COUPLES' ATTRIBUTIONS FOR WORK FUNCTION CHANGES IN PRODROMAL HUNTINGTON DISEASE

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Abstract

People who have tested positive for the expanded Huntington disease (HD) gene who are not yet diagnosed (pre-HD) and their companions report subtle changes in ability of people with pre-HD to do their jobs. However, it is not known whether they attribute these changes to HD. Semi-structured telephone interviews were conducted with seven people with pre-HD at different estimated points from diagnosis and six companions. Data were analyzed using qualitative analysis methods.

Participants made attributions related to health, work, and temperament. Only one participant attributed a change to HD. The process of forming attributions was demonstrated through symptom monitoring and comparison of participants with pre-HD to others with and without HD. Participants also expressed uncertainty regarding how to make attributions.

Attributions influence coping procedures, including whether to seek and accept medical treatment. In people with prodromal HD the relationship between attributions and use of coping strategies for symptoms that interfere with job functioning is unknown.

Keywords Huntington disease; Common Sense Model; qualitative research

CHAPTER 2

COUPLES' ATTRIBUTIONS FOR WORK FUNCTION CHANGES IN PRODROMAL HUNTINGTON DISEASE

Introduction

When people notice changes in their functional abilities, such as memory problems or fatigue, they try to find explanations for these changes. The attributions they choose to explain these changes influence how they cope with these changes. Leventhal and colleagues created the Common Sense Model of Illness Representation to describe this phenomenon (H. Leventhal et al., 1980). The model is based on the observation that when people perceive somatic changes it is common sense to assign attributions to them and to select appropriate coping procedures based on theses attributions (H. Leventhal et al., 1998). The Common Sense Model has been used widely to describe how people form illness representations, also called illness perceptions. The Common Sense Model has been used extensively to describe illness representations in people with multiple sclerosis (Lerdal, Celius, & Moum, 2009), heart failure (Jurgens, Hoke, Byrnes, & Riegel, 2009), hypertension (Chen, Tsai, & Lee, 2009), and myocardial infarction (Broadbent, Ellis, Thomas, Gamble, & Petrie, 2009b).In the present analysis, the Common Sense Model is used to explore illness representations in HD gene-expanded individuals and their companions in the preclinical stages of HD.

Huntington disease is an autosomal dominant neurodegenerative disease characterized by progressive motor, cognitive, and behavioral decline. Age of diagnosis is based on classic motor symptoms and is typically during middle age; death occurs approximately 17–20 years after diagnosis (R. H. Myers, 2004). The disease involves a trinucleotide (CAG) expansion of the huntingtin gene on chromosome 4p16.3 (OMIM, 2010b). Prevalence of HD in North America is approximately 1 in 10,000 individuals (F. O. Walker & Raymond, 2004). Until recently, the period prior to clinical diagnosis has

been referred to as “presymptomatic.” However, current data suggest this term is a misnomer since functional changes prior to clinical diagnosis may represent symptoms of impending HD; thus, a more appropriate term for the period prior to clinical diagnosis is “prodromal” (Paulsen, 2010).

According to the Common Sense Model, assigning attributions is an active process, beginning with an appraisal stage in which symptoms are first noticed. At this point symptoms may be subtle and nonspecific. The appraisal stage may be longer for symptoms that are mild or ambiguous and may result in making benign attributions instead of threatening ones (H. Leventhal et al., 1998). As symptoms progress, they may become more severe and distinctive, at which time people are more likely to attribute them to illness. The active process of assigning attributions to perceived changes involves both emotional and cognitive processes and is based on the “symmetry rule”: People seek labels for perceived symptoms; given labels, people seek symptoms to confirm the labels (H. Leventhal et al., 1998). This phenomenon was demonstrated in a study in which students with normal blood pressure, when told they had hypertension, proceeded to identify symptoms of high blood pressure such as headaches, palpitations, tension, dizziness, and flushing (Baumann et al., 1989).

People form illness identities based on information from three sources: perceived symptoms, external sources (including healthcare providers, family, and media), and past experience with the illness (H. Leventhal et al., 1984). Past experiences with an illness may lead to inaccurate illness attributions. For example, people who have had past experience with influenza, cancer, hypertension, depression, or schizophrenia, either in themselves or a close family member, held illness beliefs that were not always consistent with medical knowledge (Godoy-Izquierdo, Lopez-Chicheri, Lopez-Torrecillas, Velez, & Godoy, 2007).

Illness representations are important because they influence the coping procedures people select. Coping procedures are defined as “the cognitive and behavioral actions we

take (or do not take) to enhance health and to prevent, treat…and rehabilitate from illness" (H. Leventhal et al., 1998, p. 722). This includes whether to seek medical care and accept treatment.

A group of researchers used the CSM to explore illness perception in people diagnosed with HD and their spouses. In one study, patients' acceptance of HD was positively related to their mental health (Helder, et al., 2001). The authors maintain that acceptance allows people with serious illnesses to use more effective coping mechanisms. In another study (Kaptein, et al., 2007), people with HD and their spouses had similar illness representations; however, spouses reported more symptoms than people with HD and attributed more symptoms to HD. People with HD attributed many of their symptoms to stress. Also, spouses who believed less in a cure for HD had a more positive impact on their partners' vitality and social functioning. At the same time, people with HD who had a stronger belief in control over their illness reported a higher quality of life. In another study, spouses seldom used denial as a coping method, and denial decreased as the severity and duration of HD symptoms increased (Helder, et al., 2002). In total, these studies indicate that acceptance of HD is related to better outcomes and it is not necessary, or even desirable for members of a couple to have the same illness representations.

The studies discussed above were conducted by the same team of researchers and involved people already diagnosed with HD. No data could be found that explore illness representations in prodromal HD. It has been possible for people at risk for HD to receive definitive genetic testing to determine whether they will develop HD since the gene expansion was discovered in 1993 (The Huntington's Disease Collaborative, 1993). While clinical diagnosis of HD is not made until a person displays distinctive motor signs (e.g., chorea, dystonia, and impaired voluntary movements) (Hogarth, 2003), recent research indicates subtle changes in cognition, behavior, and motor control can appear years before clinical diagnosis (Paulsen et al., 2007). Neuroimaging studies indicate

structural and functional changes in the brain also occur prior to clinical diagnosis (Kloppel et al., 2009; Paulsen, 2009; Paulsen et al., 2007).

The changes that occur in prodromal HD may affect a person's ability to maintain day to day functions, including work functions. Family members may be the first to notice functional changes; in some cases they feel as though they are the only ones who notice these changes (Williams et al., 2007). For this reason, they are useful informants in HD research. It may be especially important to include family as informants in disease processes that involve cognitive loss (Davis, 2001), as HD does. Furthermore, insight is impaired in people with clinically diagnosed HD and it is not clear at what point impairment begins (Hoth et al., 2007). This means family members may provide important information in addition to participants with pre-HD regarding functional changes in prodromal HD. Family members are also impacted by HD (Williams, et al., 2009) and therefore influence the care people with HD receive.

Family members report changes in behavior, activities, relationships, and physical abilities in HD gene-expanded individuals prior to clinical diagnosis; however, they don't necessarily know whether the changes are related to HD (Williams et al., 2007). Individuals with pre-HD and their companions also report subtle changes in work function; changes in work function may be one of the most reliable indicators of functional changes in the prodromal stages of HD (Paulsen et al., 2009).

While people in the prodromal phase of HD and their companions report subtle changes in work function, it is not clear whether they attribute these changes to HD. Attributions have consequences because they influence the coping procedures people choose, including whether to seek treatment for noticed changes. While clinicians may recognize changes in work function related to prodromal HD, it is not known whether individuals with pre-HD or their companions do. The purpose of the current analysis is to explore the attributions of people with pre-HD and their companions make for perceived changes in work function. The answer to this question may provide insight into whether

these individuals would recognize potential benefits of treatments, lifestyle adaptations, or support to maximize function and wellbeing in the prodromal period.

Methods

The data in the current analysis were obtained from semi-structured telephone interviews with individuals with pre-HD and their companions. The entire interview was part of a larger mixed methods study designed to create a tool to measure work function in prodromal HD. Work function was defined in the study as the cognitive, behavioral, and physical ability to perform expected tasks related to paid or unpaid work. The work function study is part of PREDICT-HD 2.0, a multi-site longitudinal study designed to identify and track markers of HD during the prodromal period (Paulsen et al., 2006)

Participants

Participants were identified in collaboration with the PREDICT-HD 2.0 coordinators. Purposeful criterion sampling (Sandelowski, 1995) was used to recruit individuals with HD gene expansion at different points from HD diagnosis as estimated using age and CAG-repeat number: far (≥ 15 years), mid (9–15 years), and near (≤ 9 years) (Langbehn et al., 2004). Participants are not identified in the results according to estimated proximity to diagnosis in order to preserve anonymity. A sample size of at least 6 individuals with pre-HD and six companions was considered adequate to discern meaningful results from the qualitative interviews (Morse, 1994).

Nine individuals with pre-HD and eight companions agreed to particate—four individuals with pre-HD were classified as far from diagnosis, two midway from diagnosis, and three near diagnosis. Two participants with pre-HD were later identified as having received a clinical diagnosis of HD since their last classification and were excluded from the current analysis. Age range of participants with pre-HD was 30–59 years; median age was 44.5 years. Six participants with pre-HD were female. All

companions were either spouses or significant others of participants with pre-HD. The university Institutional Review Board approved the study.

Procedure

The interviewer (NRD) contacted prospective participants by telephone and asked them whether they were willing to participate in a study about changes in work function in "presymtpomatic HD." After participants returned mailed consent documents, the interviewer contacted them for semi-structured telephone interviews at a pre-arranged time. Participants responded to questions regarding changes in individuals' with pre-HD work function including changes in physical skills, keeping track of information, getting places on time, relationships at work, quality of work, mood, and interest in work (see Table 1 for sample questions). The interviewer also asked participants to describe how individuals with pre-HD manage changes in work function and whether they have advice regarding work function for others with presymptomatic HD. The interviewer asked participants to comment on any other issues related to work function that were not asked in the interview. The interview did not include questions regarding attributions for noticed changes. However, most participants volunteered attributions. Interview length varied according to how many changes in work function participants described and continued until participants had nothing more to add. Average interview length was 26 minutes.

Data Analysis

Data were analyzed using qualitative descriptive techniques (Sandelowski, 2000). One coder (NRD) initially coded the data using NVivo8 software (QSR International, 2000). The coder did not use a conceptual framework during the initial analysis; rather, the theme "attribution" was identified in the data. Following the identification of the attribution theme, the initial coder conducted a literature search on attribution and selected the Common Sense Model as a framework for further data analysis. An expert in

HD research and a researcher who uses the Common Sense Model in healthcare research confirmed the appropriateness of using the model in this context. The initial coder then returned to the data and coded again looking for themes related to the Common Sense Model. The study was not designed using the Common Sense Model; hence, there were no data in the interviews directly related to attributes of time-line, consequences, control, or cause. However, the coder found data related to attributions as well as the active process of making attributions for noticed functional changes. Codes were created for attributions and for active processes.

After coding the data using the Common Sense Model framework, another member of the research team (JKW), an expert in HD, assessed reliability of the coding. The two investigators discussed differences until 100% agreement was reached. The remaining members of the research team assisted in assessing the validity of the data analysis conducted by the initial coder and the HD expert. The entire team discussed validity issues including: definition of coding terms, clarification of themes, descriptive validity (the themes identified by the initial coder and verified by the expert were apparent in the data to the other team members), and interpretive validity (the interpretations of the data made by the initial coder and the expert made sense to the other team members) (Sandelowski, 2000). Discussion continued until 100% agreement was reached.

Results

Most participants volunteered attributions for noticed changes in work function. Although participants were asked specifically about work function, several talked about changes at home as well, including irritability and memory issues. Some participants may have been influenced by external illness labels when making attributions. For example, a companion stated, “Just by the nature of it being ‘presymptomatic,’ it doesn’t seem to be affecting her yet.” On the other hand, consistent with the Common Sense Model

symmetry rule, a participant with pre-HD expressed the tendency to attribute changes to HD based on the label: “So you don’t know if it’s the Huntington’s or just knowing that you’re positive for Huntington’s?”

Most participants, however, did not attribute changes to HD. Three categories of attributions for reported changes in work function were identified: health-related, work-related, and temperament-related. In addition, participants also provided insight into some of the active processes they used to make, or try to make, attributions for noticed functional changes. Four themes related to active processes were identified: symptom monitoring, comparison to others with HD, comparison to others without HD, and comparison to past self. Participants also expressed uncertainty regarding how to make attributions for noticeable changes in work function.

Attributions

Health-Related Attributions

Health-related attributions for functional changes were common among both participants with pre-HD and companions. However, only one companion attributed a spouse’s functional changes to HD. Several other health-related attributions were mentioned, with aging being the most common.

Aging

After discussing subtle changes in memory, physical ability, or relating to others in the work environment, participants indicated they attributed many noticed changes to aging. For the most part, individuals with pre-HD minimized or normalized these changes, indicating they were not considered very severe: “Oh my gosh, [it’s] not the beginning of the end, it’s just getting old”; “So, normal little age quirks that show up [that] you notice with one’s body when you’re 30 and 40 and 50.” Some noticed changes that were more specific and potentially more severe, but they nevertheless attributed them

to aging. [Referring to coworkers:] “I look around at people my age and…you’ve known so many people that have come and gone; after awhile it kind of seems to be kind of a numbness—you don’t get engaged to people.”

Companions also attributed functional changes to aging, including increased use of visual reminders such as “to-do” lists and decreased socializing at work. Some companions normalized partners’ memory problems by attributing them to aging, which affects everyone. “[O]ur memory is not the same as when we are in our fifties as it was in our forties or thirties.”

Other Health-Related Issues

Several participants with pre-HD and companions offered other health-related attributions for noticed functional changes including allergies, ankle and wrist injuries, accidents, menopause, and other health conditions. Some drew a distinction between physical changes due to injuries versus those that may be attributed to HD. “I have had injuries. I lost a ligament in my right foot…. I broke my left wrist skiing last winter, so it’s a little stiff….Nothing from Huntington’s.” Even when behavior was notably different it was not attributed to HD: “I would say she has gotten moodier….she’s having a terrible menopause. You know, versus when she was younger….If I had to put my finger on one definitive cause…I would think that would be it.”

Work-Related Attributions

Participants with pre-HD made attributions for changes in their work function related to their work situation, including problems with supervisors or coworkers, having a bad day, and being overworked: “I got my first negative review ever this year, that I ever had in my life, but then I asked around and I found out that they gave every single person a negative review”; “I used to remember stuff all the time. But, I didn’t have so much on my plate….I didn’t finish [an assignment] on time. And, I don’t know if that’s just because I have so many things going on….” In fact, if a participant with pre-HD

mentioned HD, the person disclaimed it as an attribution: "[S]omebody may say, 'Well, that is just Huntington's'; and it's not Huntington's….The guy having a scream attack because he is having a bad day is just okay."

Temperament-Related Attributions

The behavior of many of the individuals with pre-HD was described in terms of their pre-existing personalities. Most of the time problems noted in work function were not considered changes, but rather a reflection of their temperaments: "No changes; I am chronically late"; "I've never been a super coordinated person anyway." Again, if a companion mentioned HD, often it was to deny it as an attribution:

> She being a perfectionist, certain things really piss her off….Like, she's obsessively clean and neat; so if you bring a leaf in, you better take it out. So I don't think it has nothing to do with Huntington's. It's just being a witch [laughs].

Even when companions acknowledged behavior at work had changed, they attributed it to temperament. "[s]he gets much more stressed out, like, easier… [but] she has always been high strung." Participants with pre-HD also attributed change in behavior at work to a change in temperament:

> I definitely have a lot more confidence when it comes to arguing [at work]. Before I think I would have swept it under the rug…and now I'll just battle with them….I've gotten the confidence to be sort of obnoxious.

Active Processing

According to the Common Sense Model, forming illness representations is an active process involving both cognitive and emotional pathways. Participants in this study provided insight into the active processes they used for evaluating changes they noticed in their or their partners' work function abilities. Participants displayed active processing through monitoring symptoms and comparing individuals with pre-HD to

others with and without HD. Not all active processes resulted in an attribution. Several participants expressed uncertainty regarding the meaning of some functional changes.

Symptom Monitoring

A few participants talked about monitoring for HD symptoms. Participants with pre-HD monitored themselves: "I pay attention to things"; and companions monitored partners: "[T]he awareness is always there, yeah…. You always watch them for little tell-tale signs." Symptom monitoring included comparing current functional abilities with past functional abilities. Participants with pre-HD considered a change to be more problematic when it was "not like" their usual selves. Symptom monitoring did not necessarily result in making attributions.

Comparison to Others with HD

Participants compared changes in individuals with pre-HD work function to what they had observed in others affected by HD. Again, they did not necessarily make attributions after making comparisons. [Comparing his partner's behavior with her HD-affected father's]: "[S]he is quite stubborn and you know, her father, he was very stubborn." Some participants with pre-HD expressed reluctance to attribute changes to HD because the age of clinical diagnosis for affected parents was older than their current age: "Because Huntington's in my family tends to come later in life….I'm thinking maybe that kind of thing…may have been just too early for me to see."

Comparison to Others without HD

Participants also compared changes in function with others who do *not* have HD. In some cases participants with pre-HD attributed changes to illnesses other than HD even if comparisons didn't match very well: "…[M]y mother…has rheumatoid arthritis, but she has never had it to…this extent, and her symptoms, I guess, aren't like mine."

Companions also compared partners to themselves. “I don’t feel as good as *I* did 10 years ago either.”

Uncertainty

Several participants with pre-HD were uncertain how to make attributions for noticed changes in their work function. “I hide in my room a lot more….I don’t know why—it doesn’t make any sense. Everybody’s nice….”

> I… notice that I have periods of time when I ‘ruminate?’ I can’t shake it. It can be a problem. I don’t know what that is either. I think I may have always done it; now I am becoming aware of it. But I’m not sure.

Some participants with pre-HD wondered whether changes were related to HD but were uncertain. “Sometimes I feel tired a lot….I don’t know if that is part of it [HD] or not.” Companions also expressed uncertainty:

> [I]t’s really hard to distinguish, you know, what one thing would be versus another. Every time something comes up and I think…that [HD] may be a direct cause, then I think, no,…it would have to be obvious, you know what I mean?

For some participants with pre-HD, uncertainty itself was problematic: “And so, that…frustrates me. Not knowing.”

> I think a lot of it is…the uncertainty of what is going to happen in the future. Because, at some point, it *is* going to start to affect me. And at some point it is going to start to affect my ability to do my job. And every now and then I think about that, and you know, it worries me.

Although participants were not specifically asked to do so, they made attributions for changes they had noticed in individuals with pre-HD work function. Participants also demonstrated the use of active processes in either deciding or not deciding to make attributions for noted functional changes. Participants expressed discomfort with uncertainty regarding how to make attributions for noted changes and uncertainty regarding when changes will affect their ability to do their jobs.

Discussion

The findings in this analysis provide preliminary evidence that people in the prodromal stage of HD and their companions notice changes in individuals with pre-HD functional abilities and make attributions for these changes; most of the time they do not attribute noticed changes to HD. The tendency to make attributions for noted functional changes illustrates the basic assumption in the Common Sense Model that people assign labels to somatic changes. According to the symmetry rule of the Common Sense Model, it would be logical to assume participants would attribute some noticed changes in work function to HD because they had been asked to participate in a research study because one member of the couple had tested positive for the HD gene expansion. Participants have previously agreed to be contacted for HD-related research. Despite this, only one participant attributed a change in his spouse's functioning to HD. In this study, most changes in work function were attributed to aging and other health conditions, work environment, and temperament. These findings appear to violate the Common Sense Model symmetry rule. This may partly be explained by the label "presymptomatic" that was used during recruitment and in the interview process. Cover letters, consent documents, and the interview script all referred to "presymptomatic HD." If people are told they or their companions are presymptomatic it may not seem logical to attribute changes to HD.

For some people at risk for HD who have not been tested, vague symptoms and uncertainty of their meaning create high levels of distress (Duncan et al., 2008). It may be beneficial for individuals with pre-HD to attribute subtle functional changes to HD if it facilitates effective coping procedures. For example, although there is no cure for HD, treatments for many symptoms exist (Adam & Jankovic, 2008). Some treatments may be useful in treating early functional changes noted in this study and others, including medications to improve mood, ability to focus (Grimbergen & Roos, 2003), and irritability (Ranen, Lipsey, Treisman, & Ross, 1996). These symptoms potentially

interfere with important aspects of quality of life such as work and relationships. Evidence of structural brain changes prior to HD diagnosis suggests it may be important to target affected brain regions with medications prior to significant structural changes (Hannan, 2005). Attributing functional changes to HD prior to diagnosis may also allow individuals with pre-HD and their companions more time to plan for the future, including rearranging family roles.

Of course, there are ethical considerations in attributing subtle functional changes to HD prior to diagnosis. These include risk of psychological harm (Bloch, Adam, Wiggins, Huggins, & Hayden, 1992; Witjes-Ane et al., 2007) and exposing individuals with pre-HD and their families to discrimination, both identified as important issues for people in HD families (Penziner et al., 2008). The results of the current analysis do not reveal whether making HD attributions for functional changes in prodromal HD has a positive or a negative impact on coping or quality of life. The data in the current analysis were collected in the context of evaluating work function and were not designed specifically to identify participants' attributions for perceived functional changes or how attributions influenced coping procedure selection.

In the current analysis, only one spouse attributed a symptom (choking on food and pills) to HD. While this was an exceptional finding; choking is a distinctive, not a vague, symptom, and thus difficult to assign to other, more benign, attributions. Most of the functional changes reported were vague, mild, and ambiguous, to which people tend to make benign, non-threatening attributions (H. Leventhal et al., 1998). At the same time, it is important to keep in mind that the subtle functional changes noted by participants in the current study could indeed be attributed to labels other than HD. After all, people who are gene positive for HD also age, and they have other illnesses and circumstances that may contribute to noticeable changes in work function. Family members have expressed frustration when healthcare providers do not adequately address health issues other than HD (Williams et al., 2007). On the other hand, recent research

indicates changes that occur prior to diagnosis have the potential to impact work function (Paulsen et al., 2009).

Curiously, many of the attributions that were stated by participants were attributions over which participants have little or no control, such as aging and temperament. One may assume people faced with an uncontrollable disease would select attributions over which they had more control, but this did not seem to be the case. On the other hand, aging and temperament may be seen as "normal" phenomena which may decrease the perception of their severity. These are examples of normalization and social comparison which are both common in illness representation (H. Leventhal et al., 1980; H. Leventhal, E. A. Leventhal, & Nguyen, 1985). Normalization involves attributing changes to non life-threatening causes or using social comparison to attribute changes to things "normal" people experience. This has been noted in older people who compare their physical changes to others and attribute them to aging, even when they may actually be related to illness (E. A. Leventhal, 1984). The comparison to others with HD is also consistent with the Common Sense Model as people tend to compare themselves to others with similar health conditions (Godoy-Izquierdo et al., 2007).

The finding that many changes were attributed to aging is not surprising given that many changes were mild and ambiguous. Aging is related to subtle functional changes, including mild cognitive decline (Caserta et al., 2009). Aging is also related to physical decline and decreased social activities (Buchman et al., 2009). People who are middle-aged are more likely than older people to use avoidance coping procedures and delay seeking medical care when symptoms are ambiguous (E. A. Leventhal, H. Leventhal, Schaefer, & Easterling, 1993). This could have consequences for prodromal HD; if people attribute changes to aging they may be less likely to seek medical treatment that could potentially improve their functional abilities.

In spite of the proposed usefulness of denial as a coping procedure, there is evidence that acceptance of illness by people with HD may be more effective, while

avoidance coping procedures (including denial and minimizing through normalization) have a negative effect on well-being (Helder, Kaptein, Van Kempen, Weinman, Van Houwelingen et al., 2002a; Helder, Kaptein, Van Kempen, Weinman, Van Houwelingen et al., 2002b). Acceptance of illness is associated with the use of more effective coping procedures in women with breast cancer (Carver et al., 1993). In the current study, there were few data regarding the coping procedures participants selected based on their attributions for functional changes. Therefore, further research is needed to determine whether making HD attributions for functional changes in prodromal HD results in use of coping procedures that enhance quality of life.

In the Common Sense Model, representations of the self often overlap with illness representations (H. Leventhal, Kelly, & E. A. Leventhal, 1999). Thus, the untangling of temperament and subtle functional changes related to prodromal HD may be complicated. It is possible that only when one confronts a change that is "not like" the individual with pre-HD does one begin to question prior attributions.

While participants provided insight into the active processes they used to make attributions, not all active processes resulted in illness attributions. This may not be surprising given that most changes were vague and ambiguous. However, active processing may lead to an increase in HD attributions as functional changes become more distinct and severe.

Participants with pre-HD and their companions felt uncertainty regarding how to make attributions for noticed functional changes and whether these changes were related to HD. This finding is important because uncertainty is problematic for some people with pre-HD and their family members. Some people who have tested positive for the HD gene expansion undergo testing to relieve uncertainty (Duncan et al., 2008). Furthermore, some at-risk individuals undergo confirmative testing only after they suspect functional changes are symptoms of HD (Holt, 2006), indicating a desire for certainty. Family members also report distress due to uncertainty regarding the meaning

of changes in prodromal HD and state a desire for more information (Williams et al., 2007). On the other hand, those who undergo testing to relieve uncertainty may experience more psychological distress before and after testing than those who cite specific reasons for testing (Decruyenaere et al., 2003).

Individuals with pre-HD and their companions may benefit from more education from healthcare providers regarding what changes to expect prior to HD diagnosis and what treatments are available to help maintain functional abilities longer. Some people who consider themselves or their partners "presymtomatic" will not attribute changes to HD. This supports the literature that suggests labels given by healthcare providers influence illness representations. Changing patients' illness perceptions in some disease processes such as diabetes (Lawson & Harvey, 2009) and myocardial infarction (Petrie et al., 2002) may improve health outcomes by facilitating use of more effective coping procedures. Thus, if healthcare providers inform individuals with pre-HD and their companions that subtle functional changes may be related to HD, it is possible they will attribute some changes to HD. The term "presymptomatic" should be avoided as it creates the misconception that there are no symptoms prior to HD diagnosis.

In the future it may become critical to determine the best time for therapeutic intervention in prodromal HD if interventions can delay structural brain changes associated with functional decline (Georgiou-Karistianis et al., 2003). The current analysis indicates the importance of clarifying when symptoms of HD actually begin and sorting out changes that are related to prodromal HD versus other factors. If individuals with pre-HD and their companions do not recognize that functional changes are related to HD, they may miss opportunities for interventions to treat or delay onset of symptoms that interfere with function and negatively impact quality of life.

Furthermore, acceptance of chronic illness and relinquishing control may be empowering for some people (Aujoulat et al., 2008). Patients who are uncertain how to make attributions for noticed physical changes have expressed dissatisfaction with

healthcare provider interactions (Frostholm et al., 2005). It may be possible for healthcare providers to improve communication with patients by discussing their illness perceptions with them (de Ridder et al., 2007). Therefore, people in the prodromal stages of HD may benefit from direct discussions with healthcare providers regarding their illness perceptions, including how they make attributions for functional changes. They may also benefit from more education that addresses changes they may expect in their functioning prior to diagnosis in order to reduce uncertainty.

The results of this analysis may only apply to people who have chosen to have the predictive test for HD. People who undergo predictive HD testing may use denial and minimization less frequently than those who forgo testing (Decruyenaere et al., 2003). Couples who choose to undergo testing may also have more positive relationships prior to testing (Quaid & Wesson, 1995). Thus, the subgroup of people who undergo predictive testing may share certain characteristics, and the results presented here may not apply to at-risk people who forgo testing.

Conclusions

Individuals with pre-HD and their companions make attributions for perceived changes in work function in prodromal HD. Most do not attribute changes to HD. Although some participants in the current study talked about how they manage changes in work function, not enough data were available regarding how individuals with pre-HD and their companions select coping procedures based on the attributions they made. Further research is necessary to determine whether attributing functional changes to HD in prodromal HD results in the selection of effective coping procedures and how coping procedures impact quality of life for both individuals with pre-HD and their companions.

Table 1. Sample of Semi-Structured Interview Questions

What kinds of physical skills do you use in your job? What changes have you noticed?
How are things going when you need to keep track of information?
What about getting along with other people at work?
People sometimes say that people with presymptomatic HD don't feel as well as they used to, and that can make it more difficult to do their jobs. What is your experience?

CHAPTER 3

ILLNESS REPRESENTATIONS IN PRODROMAL HD: USE OF THE COMMON SENSE MODEL IN ANTICIPATED ILLNESS

Abstract

Huntington disease (HD) is a progressive neurogenerative disease that leads to significant impairment in cognitive, behavioral, and motor function and premature death. It is caused by an expansion in the HTT gene. Testing for the gene expansion is available prior to the onset of the distinctive motor symptoms that traditionally define the onset of the disease. Recent research indicates there is a long prodromal period in HD (prHD), with changes that are detectable up to 15 years prior to onset. However, little is known regarding how persons with prHD and their companions make sense of noticed changes. The Common Sense Model (CSM) was developed to explore illness representations. The CSM is ostensibly useful in exploring anticipated illness. The purposes of this study were to explore illness representations in persons with prHD and their companions using the CSM as a framework and evaluate the usefulness of the CSM in anticipated illness using prHD as a model. Twenty-three couples participated. Results indicated participants noticed changes, made attributions for changes, used coping strategies, and evaluated them, thus using elements of the CSM. However, participants did not use all of the elements all the time, suggesting illness representation in prHD is "in progress." Furthermore, there was only partial support for other tenets of the model. However, results provide information regarding possible interventions that could be used to help persons with prHD and their companions cope with changes. Thus, the CSM was useful in describing illness representation in prHD and may be useful in other anticipated illnesses.

Introduction

Persons who have tested positive for the Huntington disease (HD) gene expansion before diagnosis are usually healthy but live in anticipation of functional decline and eventual disability. Huntington disease is autosomal dominant, meaning that offspring of persons with the gene expansion have a 50% risk of inheriting the disease (R. H. Myers, 2004). Prevalence of HD in the United States is approximately 1 in 10,000 (Walker, 2007). This means there are approximately 30,000 people diagnosed with HD in the US and another 150,000 people living at risk (NINDS (National Institute of Neurological Disorders and Stroke), 2009). Direct testing for the gene expansion has been possible since 1993 and is based on the number of trinucleotide (CAG) repeats an at-risk individual has on the HTT gene on chromosome 4 (OMIM, 2010b). Average age of onset of distinctive motor symptoms is between 35-55 years (Quarrell, 2008) and is related in part to individuals' CAG repeat length (Langebehn, et al., 2004). Huntington disease develops in persons with 36 or more CAG repeats (Quarrell, 2008).

The CAG expansion on the HTT gene leads to the creation of abnormal huntingtin protein, which causes the premature neuronal cell death associated with HD symptoms: abnormal motor movements; behavioral and mood changes such as apathy, irritability and depression; and cognitive changes, including impaired memory and executive functioning (Walker, 2007). Symptoms become progressively worse and death occurs approximately 17-20 years after onset (R. H. Myers, 2004). Common causes of death include pneumonia and aspiration, cardiovascular disease, cachexia, suicide, and accidents (Sorensen & Fenger, 1992).

Traditionally, HD onset has been defined by the presence of distinctive motor signs—chorea, dystonia, and impaired voluntary movements (Hogarth, 2003). However, a growing body of research indicates subtle changes in cognition (Stout et al., 2007), behavior (Duff et al., 2007), and motor function (Biglan et al., 2009) are detectable 15

years prior to diagnosis (Paulsen, 2010); thus, "prodromal" HD (prHD) may be a more appropriate term for the period prior to diagnosis when subtle changes begin.

While researchers are now aware that changes occur long before onset of definitive motor signs, it is not clear whether persons with prHD or their companions are aware of this. In fact, very little is known regarding whether persons with prHD and their companions notice changes or attribute them to HD. Preliminary findings presented in Chapter 2 suggest they notice changes but they do not attribute them to HD (Downing, Wiliams, & Paulsen, 2010). Rather, they attribute them to things "normal" people experience, such as aging, temperament, or other health conditions.

Illness representations are important because they influence the type of coping procedures people use; these, in turn, impact quality of life (H. Leventhal et al., 1998). It is possible to alter illness representations, and thus potentially improve quality of life by facilitating more effective coping procedures (de Ridder et al., 2007; Rozema, Vollink, & Lechner, 2009). The purposes of the present study were to explore illness representations in persons with prHD and their companions using the Common Sense Model of Illness Representation (CSM: H. Leventhal et al., 1998) as a framework and evaluate whether the CSM is a useful theoretical framework in prHD. The research questions included: 1) Do persons with prHD and their companions notice changes in daily functioning? 2) Do they form illness representations related to these changes? 3) Do they use coping strategies to address changes? 4) Do they evaluate coping strategies? 5) Do they reappraise their illness strategies based on the effectiveness of coping strategies? The results of this study may provide insight into how persons with prHD and their companions experience the HD prodrome. These findings may help identify useful interventions to facilitate effective coping in persons with prHD and their companions. Results also provide information regarding whether the CSM is a useful framework in anticipated illness.

Theoretical Framework

According to the CSM, when people receive health relevant information they form illness representations. Forming an illness representation is an active process that includes noticing somatic changes, forming an illness representation, selecting coping procedures, and evaluating the coping procedures (H. Leventhal et al., 1998). Illness representations consist of five dimensions: identity, time-line, consequences, cause, and controllability (H. Leventhal et al., 1998). "Identity" is comprised of the symptoms and labels people use to form an illness identity. Labels may come from a healthcare provider's diagnosis or from the individual trying to make meaning of perceived somatic changes. Causes refer to beliefs about what effectuated changes. Timeline refers to beliefs about the course of the illness, whether it is acute or chronic, constant or episodic. Consequences refer to beliefs about the severity of the illness and its impact on quality of life. Controllability refers to beliefs about how much influence individuals have over an illness and whether there are effective treatments and/or cures.

Forming an illness representation is an active, "parallel" process (H. Leventhal et al., 1984, p. 220) that involves both cognitive and emotional pathways. Coping strategies are "the cognitive and behavioral actions we take (or do not take) to enhance health and to prevent, treat…and rehabilitate from illness" (Lazarus, 1998, p. 722). The illness representation process is recursive; i.e., if the coping measures are evaluated to be ineffective, the illness identity is altered and/or different coping measures are selected and reevaluated (H. Leventhal et al., 1984). If coping measures are effective, this is presumed to enhance health-related quality of life (Lazarus & Folkman, 1984). Thus, coping acts as a mediator or moderator between illness representations and quality of life.

According to the CSM, not only do people label symptoms, but they also seek symptoms to match illness labels. This is known as the symmetry rule (H. Leventhal et al., 1998). Given the symmetry rule, one may assume that persons who have tested positive for the HD gene expansion would search for HD symptoms and attribute

perceived changes to HD. However, preliminary evidence does not support this. While family members noticed changes in persons with prHD, including irritability, impaired judgment, sleep disruption, balance problems, and relationship problems, they were unsure whether changes were related to HD (Williams et al., 2007). In Chapter 2, persons with prHD and their companions noticed subtle changes in work function but rarely attributed them to HD (Downing, Williams, & Paulsen, 2010). Most common attributions for changes were aging, temperament, and other health conditions. These findings seem to violate the symmetry rule. However, the results of Chapter 2 were considered preliminary due to the following limitations: small sample size (N=13), participants were only asked about changes in work function, and they were not asked to make attributions for noticed changes. The fact that most participants volunteered attributions supports the first part of the symmetry rule— that people seek labels for perceived somatic changes. The use of the term "presymptomatic" in that study may have introduced a label that inadvertently cued participants to believe changes could not be related to HD. In fact, a spouse stated he was not expecting symptoms since his wife was "presymptomatic."

It is important to include companions in HD-related research because not only are they also affected when their partners experience changes (McCabe et al., 2008) but companions may be the first to notice them (Williams et al., 2007). They also may notice more symptoms than their partners (Kaptein et al., 2007). Changes in cognitive function in persons with prHD may also make collection of collateral information from family members important. Insight has been demonstrated to be impaired in persons with prHD (Duff et al., 2010a). Also, companions form their own representations of HD and use different coping procedures than their spouses (Kaptein et al., 2007). They may influence how their partners cope, including whether they seek medical advice or accept treatment for changes. Companions often become caregivers when their partners' symptoms get severe (Roscoe, Corsentino, Watkins, McCall, & Sanchez-Ramos, 2009) and they worry about their children's risk of inheriting the HD gene expansion (Williams et al., 2009).

For these reasons, it is important to understand how companions of persons with prHD form illness representations and how they cope with partners' functional changes.

The authors of the CSM state the model can be applied to anticipated illness (Brownlee et al., 2000); beyond this assertion they do not describe how to do so, and they concede this has not been adequately tested. One group of researchers (van Oostrom et al., 2007) applied the CSM to anticipated genetic illness in persons undergoing presymptomatic breast cancer or colorectal cancer. However, their results were related to participants' representations of the illnesses and did not address their perceptions of their current levels of functioning.

Another potential limitation of using the CSM to describe illness representation in persons with prHD is that functional changes are usually subtle and people don't consider themselves ill, *per se*. Another important question is at what point do perceived functional changes become symptoms of illness? People tend to attribute subtle and ambiguous changes to things that are benign and less threatening and wait until changes are more distinctive and severe before attributing them to illness (H. Leventhal et al., 1998). Many changes in prHD are subtle and ambiguous, which may help explain why participants attributed noticed changes to things other than HD (Downing et al., 2010). In that study, participants demonstrated they used active processes to form representations by comparing persons with prHD to others with and without HD; however, they did not always arrive at an attribution for these changes.

Evaluation of the CSM

Hagger and Orbell (2003) conducted a meta-analysis of 45 studies using the CSM from 1977-2002. They looked for evidence that illness representations were related to coping behaviors and health outcomes, as the authors of the CSM assert. They found that belief in severe consequences of illness and a strong illness identity (operationalized as the presence of many symptoms) were both mildly correlated with the coping strategies

avoidance/denial and expressing emotions; belief in controllability and curability of illness was mildly correlated with generic problem-focused coping (any active attempt to address the problem). All correlations were small (0.21-0.27); however, they did not test for mediation or moderation effects, which might mask the relationship between illness representations and coping behaviors. With regard to the relationship between illness representations and health outcomes, they found that a belief in severe consequences of illness was moderately correlated with higher psychological distress and lower psychological well-being, role functioning, social functioning, and vitality. A strong illness identity was mildly correlated with increased psychological distress. Belief in a long timeline of illness was mildly correlated with increased psychological distress. Thus they conclude there is some support for the validity of the CSM.

The CSM in HD Research

There are two studies using the CSM that explored illness representations of persons with diagnosed HD and their spouses published after Hagger and Obell's meta-analysis. Both persons with HD (Helder, Kaptein, Van Kempen, Weinman, Van Houwelingen et al., 2002a) and their spouses (Helder, Kaptein, Van Kempen, Weinman, Van Houwelingen et al., 2002b) reported a strong HD illness identity (high number of symptoms), severe consequences, little hope for a cure, and a genetic cause. Spouses were more certain than persons with HD that symptoms could be controlled. Persons with HD and spouses both used acceptance, active coping, and planning coping strategies more frequently than other strategies. The researchers used correlation and regression analyses to explore relationships between elements of the CSM: illness representations and coping; illness representations and health outcomes; and coping and outcomes. They found mild to moderate relationships between these variables but they did not test for mediation or moderation effects of coping strategies.

Significance

There are several reasons why it is important to understand the illness representations of persons with prHD and their companions. First of all, illness representations influence how people cope, including whether or not they seek medical advice or accept treatment. Attributions are especially relevant in selecting coping procedures. For example, women were less likely than men to seek timely treatment for myocardial infarction because they didn't attribute symptoms to heart attack (Martin et al., 2004). People may also adhere better to treatment if their illness representations are congruent with their healthcare providers' assessment of their condition (Iacoviello et al., 2007).

Illness representations are highly individual and influenced by personal, environmental, social, and cultural contexts (Diefenbach & Leventhal, 1996). Personal factors include past experiences with an illness which may not be consistent with current medical knowledge (H. Leventhal & Colman, 1997). For example, persons with hypertension who believed that blood pressure is labile reported more lability in response to a stressor than persons who believed that blood pressure is fairly stable (Baumann et al., 1989).

Finally, if healthcare providers explore patients' illness representations, it may positively impact patient outcomes. Patients expressed greater satisfaction when healthcare providers asked them about their illness representations (de Ridder et al., 2007). Since healthcare providers can influence illness representations, it may be possible to alter inaccurate illness representations and facilitate more effective ways of coping. Interventions to alter illness representations have improved treatment adherence in persons with diabetes (Lawson & Harvey, 2009) and reduced stress in persons who had myocardial infarctions as well as their spouses (Petrie et al., 2002).

However, there is also a risk of harm in challenging persons' illness representations; it should only be attempted if it has the potential to result in more

effective coping mechanisms (Shiloh, 2006). Thus, it is important to sort out what changes in prHD may be related to HD and which are related to other factors in order to select appropriate interventions. It is important to intervene when functional changes interfere with the ability to carry out activities of daily living, such as taking care of children or maintaining employment. Behavioral changes, including moodiness, anger, and irritability, are particularly distressful for family members of persons with prHD (Williams et al., 2007). Young adults who were raised by a parent who later was diagnosed with HD reported higher levels of family dysfunction than a comparison sample (Vamos, Hambridge, Edwards, & Conaghan, 2007). Changes in prHD may also impact work function (Paulsen et al., 2009; Paulsen, Wang et al., 2010). Unemployment has been related to greater psychological distress in persons with prHD (Almqvist, Bloch, Brinkman, Craufurd, & Hayden, 1999). Intervening before the onset of definitive motor signs may be critical (Hannan, 2005) because brain imaging studies indicate that by the time of HD diagnosis, over 50% of neuronal cell death has already occurred (Aylward, 2007). Some behavior changes that occur in prHD may respond to treatment (Phillips et al., 2008; van Duijn, 2010). Therefore, awareness of changes that have the potential to interfere with day-to-day functions in prHD may facilitate interventions to help maintain function longer and positively impact wellbeing.

Methods

Design

The data presented in this paper are part of a larger, mixed-methods study designed to explore illness representations and coping in prHD. Results of the mixed methods coping analyses are presented in Chapter 4. The aims of the present chapter were to explore illness representations in persons with prHD and their companions and evaluate the use of the CSM in anticipated illness using prHD as a model. Thus, the design is qualitative descriptive. The authors of the CSM have stated qualitative

methodology is necessary to adequately capture all the dimensions of the CSM (Diefenbach & H. Leventhal, 1996). Although a theoretical framework (the CSM) was used, the purpose of the framework was not to form *a priori* hypotheses, but rather to provide a systematic way of exploring the process of illness formation in anticipated illness.

Sample

The recruitment goal for this study was 30 couples. Morse (2000) stated 30-50 participants are necessary to obtain meaningful data when using semi-structured interviews because responses lack the depth of other qualitative methods. Walter and Emery (2006), for example, reached data saturation after 30 semi-structured interviews that explored illness representations of persons from families with familial diseases. Other researchers exploring illness representation in persons with early stage dementia stated they reached saturation after nine semi-structured interviews (Harman & Clare, 2006). A sample size of at least 30 is also desirable for summative content analysis (Morse & Niehaus, 2009).

Participants for the current study were selected using purposeful criterion sampling methods. Purposeful criterion sampling is used to select participants who will provide rich information on the phenomenon of interest and who possess criteria of interest (Patton, 2002). Potential participants were research participants in the PREDICT-HD study. PREDICT-HD is a multi-site longitudinal study designed to identify and track markers of HD during the prodromal period (Paulsen, 2010). Persons who have independently undergone HD testing are eligible to participate in the PREDICT-HD. Persons who tested negative for the HD gene expansion serve as comparison subjects. Companions of participants also participate in PREDICT-HD to provide collateral information. Participation is done on an annual basis at 32 sites in the US, Canada, Europe, UK, and Australia. At this time, there are approximately 825 persons with prHD

enrolled in PREDICT-HD. Mean age of persons with prHD in the PREDICT-HD study is 43.95 (SD=10.33). In the current study, potential participants were identified in collaboration with the HD Center Coordinator at the University of Iowa (Anne Leserman).

Potential participants were recruited from the Iowa PREDICT-HD site and US sites that offer participants the opportunity to participate in ancillary HD-related research. There are 15 PREDICT-HD sites in the US, although not all sites offer the opportunity to participate in ancillary studies. Criteria for participation in the current study included persons with prHD who were estimated to be ≤15 years from diagnosis of distinctive HD motor symptoms based on age and CAG repeat length (Langbehn et al., 2004) and their companions. Persons >15 years from estimated diagnosis were not eligible because it was less likely they or their companions would notice HD-related changes. It was expected that more females with prHD would participate because 63.4% of participants with prHD in PREDICT-HD are female. Other inclusion criteria included ≥21 years old and English-speaking. Couples were excluded from participating if companions had severe chronic health conditions.

Procedure

The University of Iowa Institutional Review board approved this study. Persons with prHD were contacted by the researcher and asked whether they were willing to participate in a study about daily functioning and coping with noticed changes. Persons with prHD were informed it was a couples' study and therefore they would need to have a significant other who was also willing to participate. Twenty-one potential participants with prHD were identified from the Iowa site and 88 from other US sites. Prospective participants were contacted by telephone if they had a current telephone number and answered their telephones within three attempts. Those who did not have a current

telephone number or did not answer their telephones within three attempts were sent mailed recruitment information. See figure 1 for a recruitment diagram for this study.

Of the 39 persons who answered their telephones, 28 who were eligible agreed to receive mailed study information. Of these, 18 persons with prHD and their companions returned signed consents. Two couples were lost to follow-up, leaving 16 couples who were contacted initially by telephone who completed the study. Invitations were mailed to 60 prospective participants who did not have current telephone contact information or who did not answer their phones after three attempts. Only seven persons with prHD contacted by mail returned signed copies of the consent documents along with their companions; all seven couples completed the study. Thus, a total of 23 couples participated. Couples lived in 14 US states, including seven Midwest states, five Eastern states, and two Mountain states.

At the pre-arranged time, the researcher called participants and confirmed that it was still a good time for the interview before proceeding. Participants were asked to participate without their partners listening and interviews were audio-recorded with participants' consent.

Measure

Demographic information was collected prior to beginning the semi-structured interviews and included: age, gender, whether persons with prHD had children, and how long companions had known their partners. The semi-structured interview guides (Appendix A and Appendix B) were developed based on prHD literature and the elements of the CSM, and build on what was learned in the preliminary study presented in Chapter 2. The interview guides included questions regarding how things were going at home and at work, and in the social lives, physical activities, and relationships of persons with prHD. Participants were also asked about the ability of persons with prHD to make plans and perform new tasks.

After an introduction explaining the study, the interview began with a broad open-ended question. Participants were asked how things have been going in general for the person with prHD and if they have noticed any changes in physical function, behavior, mood, or thoughts. The question was intended to avoid implying that the interviewer expected functional decline and to allow participants to discuss issues that are currently most salient to them. This is important in exploratory studies because it may elicit responses the interviewer did not anticipate (Burns & Grove, 2005).

If participants mentioned problems in any areas of function, they were asked "Is this a change?" The interviewer then asked to what participants thought changes were related and how they made those decisions. At the end of the interview, participants were asked if there was anything else they had noticed that they had not yet discussed. Interviews continued until participants indicated they had nothing more to add.

Data Management

Consent documents and hardcopy interview notes were kept in a locked filing cabinet in the researcher's office. Audio-recordings of interviews were downloaded to the researcher's password-protected laptop and subsequently deleted from the audio recorder. Interviews were then transcribed by an IRB-approved transcriptionist and verified for accuracy by the researcher. All identifying data were removed during transcription and participants were referred to by number from that point forward. Word documents were imported into a password-protected qualitative data management software program, NVivo8 (QSR International, 2000) on a password-protected computer at the College of Nursing.

Data Analysis

Data were analyzed using Sandelowski's (2000) basic qualitative descriptive methodology. According to Sandelowski, basic descriptive methodology is categorical, less interpretive than other qualitative methods, and does not depend on highly abstract

analysis. The purpose is to provide "a comprehensive summary" of an experience using a preselected framework (p. 336). Descriptive validity is established when separate researchers agree that descriptions are apparent in the data. In the present study, two researchers (ND and JW) coded the transcripts of the first ten couples interviewed using the elements of the CSM as a framework.

Coding categories that represent formation of illness representations included: changes, attributions, active processes, and evaluation. These categories came from the results of the preliminary analysis which indicated participants used these categories when talking about changes. While illness representations include five dimensions—identity, timeline, cause, consequences, and controllability/cure—preliminary data did not show explicit evidence of these dimensions beyond making attributions for changes. However, the other dimensions may be implicit in the attributions people chose. The term "identity" was avoided because of the conceptual confusion created in past studies that operationalize it as frequency of symptoms and the conceptual overlap between noticed changes, identity, and causes. Thus the term "attributions" was used instead. The coders compared results and reached 100% agreement that descriptions were apparent in the data. A single researcher (ND) coded the remaining transcripts using the agreed upon coding categories and a second researcher (JW) indicated agreement with this coding.

Summative content analysis (Hsieh & Shannon, 2005) was used to report how often participants used elements of the CSM. Counting allows researchers to make judgments regarding how important findings are and to validate findings by recording how frequently they appear in the data (Miles & Huberman, 1994). Huntington disease was only counted as an attribution if participants said changes were probably or definitely related to HD.

Results

Interviews ranged from 9-58 minutes for persons with prHD (M=29), and 13-48 minutes for companions (M=25). Persons with prHD and companions noticed changes in the functioning of persons with prHD. Most participants made attributions for changes and were able to explain the active processes they used to decide what changes were changes were related to. Some discussed the active processing they used to make meaning out of noticed changes but they did not always make attributions for changes. Most participants were able to talk about coping strategies they used, although some said changes were not severe enough to warrant coping strategies at that point. For the most part, coping strategies were effective. One couple reevaluated what changes were related to based on the effectiveness of a coping strategy. Participants are identified by number following their comments: Persons with prHD are numbered P01-P23; companions are numbered C01-C23.

Changes

All participants with the exception of one married couple noticed changes in the partner with prHD. Participants noticed changes in cognitive, behavioral, and motor function. They noticed changes in their mood, relationships with partners, social life, work, and at home. For the most part, changes were subtle and did not interfere with functioning. However, in some cases, changes interfered with functioning in persons with prHD. The results of the summative content analysis of endorsed changes are presented in Table 2.

Cognitive changes were the most common, especially memory. Memory issues were mostly minor and involved an increased awareness of the need to write things down in order to remember them. Two persons with prHD mentioned difficulty remembering common words, which the companion of one also mentioned. Two companions talked

about instances where their partners had forgotten items at the grocery store. In one of these cases, the person with prHD was very upset:

> [H]e had a funny episode…where…he called me on his way home from work and said, "What do you need at the store?" and I told him….[T]hen he got to the grocery store and forgot, and called home very upset that he had forgotten….I wasn't home. When I got home later he said, "I couldn't remember what you wanted me to get, and the boys didn't know, and I was all mad at them." …[T]hat was kinda scary. [C21]

Other cognitive changes included difficulty focusing, multitasking, making plans, and doing new tasks.

Work changes included changes in employment such as early retirement, quitting jobs, unemployment and changing jobs. Two persons with prHD left their jobs—one quit to pursue a less stressful job, and one chose early retirement instead of changing positions, keeping in mind his gene status. Mood changes were also common. Three persons with prHD stated they had depression, and one had a past suicide attempt. Four companions talked about partners' depression. In two cases in which companions talked about depression, their partners with prHD did not. Other words persons with prHD used to describe their moods were: "nervous," "anxious," "stress," "panic," "not rosy," "up and down." Words companions used were: "frazzled," "worries," "moody," "stressed," and "mental issues."

Almost a third of all participants talked about irritability and impatience in persons with prHD. In addition to the words "irritated" and "impatient," persons with prHD talked about feeling "anger," "cranky," "agitated," "short-tempered," "annoyed," and "less tolerance;" companions used the words "overreacted," "grumpier," and "edgy."

More than a third of persons with prHD talked about problems in their relationships, especially fighting with their partners. Four companions talked about problems in their relationships; in all cases their partners also talked about this. Most of the fighting between couples involved verbal altercations; however, two companions said their partners had been violent—hitting them or throwing things at them. While one

person with prHD mentioned this, the other did not. The person with prHD in this couple worried about the effect on her partner:

> I know that me having this disease is hard on her….[W]e really help each other in a lot of areas….But yet when we trigger each other and are fighting, you know, then I think sometimes…maybe it'd be better for me not to be with her. [P15]

Another person with prHD said relationship problems with her husband had to do with him not accepting her HD:

> He refuses to believe I am going to get it….And that makes it hard for me because he wants you to live like you are never going to be sick….So you're not supposed to be doing anything to prepare. [P04]

Over half of persons with prHD and several companions talked about physical changes, including motor changes. Both used a variety of terms to describe physical changes: "slowing down," "tired," "clumsy," "drops" things, "off balance," "fidgeting," "little movements," "shaking," "jerky," and "twitch." Both also noticed changes in spatial judgment. For example, a proband talked about fear of using escalators, and a companion stated her partner had trouble knowing "where her body parts are." One proband talked about her "cupped" hand, one mentioned reduced strength, and one fell out of bed. Two companions noticed gait changes, including being "not as spry," and tripping when walking. A companion stated his wife's speech was slower.

Both probands and companions talked about social withdrawal, including difficulty meeting people and avoiding social interaction. One proband, for example, found social interaction tiring. Participants also talked about reduced interest in cooking and housework. Two companions noticed their partners being obsessed with activities at times and one person with prHD said she had "repetitiveness" in her head, sometimes repeating phrases over and over and avoiding certain numbers. One proband mentioned decreased sleep, while one companion noticed her partner had decreased sleep, and another that her partner slept more than previously. Two companions said their partners had decreased interest in sex.

Attributions

Participants made attributions for most changes. Many volunteered attributions without being prompted, while others were able to provide a response when asked what they thought changes were related to. Several participants stated they did not know what some changes were related to. The most common attributions were aging, temperament or personality, and other life stressors. Some mentioned other physical or health conditions such as depression or anxiety, hormones or menopause, being tired, or other illnesses. Only seven persons with prHD and seven companions attributed some changes to HD. The results of the summative content analysis of endorsed attributions are presented in Table 3. Table 4 presents the links between changes and attributions. Because attributions and active processes are connected in the CSM, more details regarding attributions are presented in the section below on active processes.

Participants most often attributed memory changes and feeling tired or slowing down to aging, although other changes were also attributed to aging: "I'm tired…I'm feeling my age" [P04]; "[She's] slowing down a bit. I think that's age, though" [C21]. Other participants attributed memory problems to stress, or being busy or "distracted."

Participants attributed several changes to temperament or personality, especially mood or irritability: "I've never been an organized person….I've always been clumsy, so that's all, you know, just part of being me" [P18] "She's moodier than most people I know, but that doesn't mean she hasn't been moody, you know, since she was born" [C22]; "[T]here are times where she can be impatient about stuff…I don't even think it's even an HD thing; I mean, it's just kind of how she is sometimes" [C03].

Life stressors were common attributions for changes, including being busy at home or at work, moving, parenting, issues with extended family members and problems with their relationships with their partners: "I don't really have friends….I'm so busy" [P19] "[S]he did have a suicide attempt….I think…she has a basic inability to handle the day-to-day life stresses of raising a family" [C20]. Some had extended family members

who recently tested positive for HD or were showing symptoms: “I have some good days and I have some bad days….[W]e recently found out that another family member has the gene” [P21]. Two persons with prHD said their depression was related to having a “crummy” [P20] or “crappy” [P12] childhood with an HD-affected parent.

Four persons with prHD attributed irritability, mood, or sleeping problems to “hormones,” two due to menopause, one due to PMS, and one due to postpartum hormonal changes. A companion stated he believed changes in the couple’s sex life were partly related to his partner’s menopause. Participants also attributed changes to other illnesses, including chronic fatigue sydrome, connective tissue disorder, and arthritis. One person fell out of bed and attributed that to arthritis and difficulty getting out of bed: “That could just happen any time” [P16].

More persons with prHD than companions attributed irritability and impatience to problems in their relationships with their partners. For most, relationship issues referred to arguing with their companions: “We fight a lot” [P01]; “[My husband] was telling me what [golf] club to use and I don’t like that” [P22]. One person with prHD talked about adjusting after recent marriage: “I actually just got remarried a year and a half ago….[H]aving someone in your space, that’s a chore” [P22]. One companion stated his partner is too focused on HD instead of enjoying life now: “[T]o her it just feels like her clock is ticking….That, to me, is what is robbing her of her joy and her quality of life” [C04]. Three couples moved recently before being interviewed and attributed some changes to the stress of moving, including difficulty meeting new people, irritability and mood changes, and issues with their relationship.

Although HD has traditionally been diagnosed based on movement changes, participants attributed movement changes to a variety of other things: “I started fidgeting a lot more….That could be my anxiety” [P04]; “[S]ometimes I will move in a way that seems involuntary….I have no way of knowing…if it was HD-related” [P05]; “I’m

guessing her gait is a little bit not as spry…[but] she's not doing as much [exercise] as she used to" [C05].

For several participants, it was difficult to make attributions for changes that were subtle and ambiguous. Several participants stated "I don't know" or "I'm not sure" when asked what they thought changes were related to. Many participants were reluctant to attribute ambiguous changes to HD: "I am so careful not to imagine it's symptoms or something, because like I say, I don't want it to happen" [P04]; "I try not to jump to Huntington's the first off" [C02]. Memory issues were particularly ambiguous: "Partly age. I don't know if it's HD-related or not" [P10]; "You know, could be Huntington's, could be that, you know, people just forget sometimes" [P20].

In four couples, both partners attributed changes to HD. In these couples, the person with prHD had several changes. Some changes interfered with functioning, including more frequent irritability, social withdrawal, work issues, and movement changes. In these cases, participants stated they were definitely changes and not related to personality or external stressors. One person with prHD was unemployed and afraid to find a new job due to difficulty multitasking and learning new things, which she attributed them HD. Her companion was frustrated because they were struggling financially. She stated her partner was afraid of having an "outburst" at work [C15].

Participants were more likely to attribute changes to HD if they were distinctive For example, one woman said her husband noticed she was holding her hand in a "cupped" way [P19]. A husband attributed his wife's "forgetfulness" to HD because it "affects her shopping" [C18]. One woman was choking while eating, a distinctive change that she attributed to "early" HD [P15].

Three persons with prHD and three companions attributed changes to HD when their partners did not. In these cases, participants also noticed several changes; however, while one person in the couple attributed them to HD, the other attributed them to other things such as personality, depression, and life stressors. One woman noticed a change

that was distinctive—being uncomfortable on escalators [P12]. Thus, she attributed it to HD. Another woman attributed her severe depression to "early onset Huntington's" [P20], while her husband thought her depression was more related to "life stressors" and the stress of being "gene positive for Huntington's" [C20]. One man was feeling "anger," which he attributed to HD because it was a change for him [P01]. His companion did not mention his anger and attributed changes she noticed in him to age. In all three cases, persons with prHD whose companions did not attribute changes to HD also stated they were having issues in their relationships with their companions.

Two companions noticed distinctive movement changes and attributed changes to HD when their partners did not. One noticed "little movements" [C17] while the other noticed "shaking" [C13]. The partner of the companion who noticed shaking stated it was from "nerves" and acknowledged her husband thought they were related to HD. The third companion who attributed changes to HD while his partner did not noticed multiple cognitive changes, including difficulty multitasking and learning new things, becoming frustrated with new tasks and fretting over planning [C18].

Active Processes

Most participants demonstrated active processes related to how they decided what changes were related to. Some paid attention to symptoms while others normalized their lives as much as possible: "I always notice changes in my behavior or things that I'm doing that might be HD related" [P05]; "I try to be, you know, fairly conscious of changes. I mean, I'm sort of always in the back of my mind expecting that there's going to be changes" [P11]. Participants made comparisons in order to decide whether changes were related to HD or to other things; they compared persons with prHD to others with and without HD, and to the way they have always been. Companions compared their partners with themselves: "So sometimes, when I see myself doing things, I'm like, 'Don't be like that; that's how your mother was!'" [P18]; "[I]t felt like I was making a

kind of face that seemed similar to what my aunt used to do" [P05]; "[H]e doesn't have any more changes, or any more difficulty remembering things than I do" [C10];

> [W]e have her mother who's in the final states now of Huntington's and we can see her. And we've seen her how she's progressed….[W]hen you get the gene from …your mother's side then typically it ends up being about close to the same…onset of symptoms [C18].

A person with prHD who was having difficulty controlling her temper said, "I've kind of always been that way… [but] it just feels a little more often and…a little quicker" [P15]. Some participants tried to figure out when HD would "start" by comparing themselves to others with HD:

> [I have] a lack of confidence in certain things…[and] when I talked to my uncle, who had this, he said that was the one thing that he noticed when he first started coming down with Huntington's is lacking his own confidence in doing things [P04];

> I drop stuff …[and] I know also from talking to my dad that that was one of the early signs, is that he would notice…himself becoming a little more clumsy [P06].

Although several participants talked about how HD started in relatives, seven participants said they did not know they were at risk for HD until they were adults. Their HD-affected parents were absent or were incorrectly diagnosed.

Participants who attributed changes to aging compared themselves or their partners with prHD to others their age. Companions sometimes compared partners to themselves: "I'd say I'm probably more…deliberate in learning new things….But again, I don't see that that's a different thing than my husband and others my age" [P12]; "[S]he feels like she has challenges with her memory….I don't think she forgets any more than I do" [C12]; "I attribute [memory issues] more to age than anything else because I notice it in myself, I notice it in the people around us" [C05];

> [M]ostly I just look for things that my friends are doing and if they…have the same problems, then I figure it's most likely age because none of them are involved in HD [P05];

> I try to sort of figure out what I think is age based on my experience of aging, you know, though I am a little younger than he is. And watching other people I know his age [C02].

Participants sometimes used feedback from others or sought out information from others regarding what changes might be related to. They read the HD newsletter [C19], "looked stuff up" [P06], and even paid attention to items being asked during their research visits: "I've asked questions to the other people in the other studies, you know, like 'Is this something normal?'" [P06];

> Sometimes I ask my husband [about changes] and he says, "Oh, you're just getting older" or "You're going through the change" [P23];

> I just met with [my psychiatrist] yesterday and her response was, well, that's the same thing that every 40- and 50-year-old comes in here and tells her [P12];

> [S]he gets frustrated easy….I know a lot of the questions that I answer every year on that HD survey does have a lot to do with patience [C12];

> [W]hen I was doing the PREDICT study, you know, there'd be one question on there I think about numbers. I go, "Wow, isn't that funny? Who knew? Numbers go through my head all the time." [P22].

Some participants tried to get information from researchers when they participated in studies:

> I'm involved in numerous studies….So I get reviewed by them all the time, and they constantly tell me that there's no…signs….I've asked them that, if you see any signs are you going to tell me? And they say "absolutely" [P11];

> He was just really stressed and really wound up and ended up taking a week off from work ….I wondered if it was maybe an HD symptom kicking in. And the people [at the research site] said, you know, there's really no way to know [C10];

Some participants normalized their situations by comparing themselves to others without HD: "I'm just like every other person that walks the face of the earth. Things change depending on what, you know, obstacles and problems come your way" [P12]; "I get my stress points like everybody else does" [P12]; "What marriage doesn't have issues?" [P20]; "[W]e all change; I call it the every-10-year change" [C07];

> I mean, there's times when anybody would maybe, could get emotional about something, or have a high or low or something. And I don't think anything that she's had like that would have been considered strange to me [C03].

Some participants talked about how the knowledge of having the HD gene expansion influenced how they thought about changes: "But obviously…you know having the HD diagnosis…[you're] always concerned" [P17]; "I mean, once you find out that you have Huntington's in the family, sometimes you might start looking for things and blaming it that way" [C23]; "[T]he thing is, when you know that someone has Huntington's, you just sort of make the assumption, okay this must be an early sign…whether it is or not" [C02].

Not all active processes resulted in making an attribution, sometimes because changes were not severe enough or weren't distinctive. Some participants were waiting for changes to be more distinctive and severe:

> I don't know….I mean, there's some of the stuff that you can't necessarily say for sure this is Huntington's….But there's some stuff that, I am sure as it comes along, I will know for sure that it's Huntington's stuff [P06];

> I don't think there's anything that I can definitely attribute to HD, although it's really hard to tell….I'm guessing it's probably going to be more genetic, more…movement, but I, again, I don't know for 100% certainty [C05];

> I'm not seeing any signs yet, and I look for them all the time. And we joke about it because we don't know whether they're signs or if it's just old age, you know?...I think what I'll see is…that he's gonna be more agitated [C07];

Participants demonstrated active processing when trying to determine whether issues were changes: "I think meeting peers has been very difficult. I can't say whether it's more or less difficult. I'd say it probably has been a little more difficult" [C20]; "I mean, she is a moody person in general,…but it doesn't seem to be as much spread between the worst of times and the best of times" [C12]. For others, issues were definitely changes: "I've never been good in mathematics, but now it's just gone to pot…it was never this bad" [P04]; "I am not as comfortable on escalators….I think that's

HD-related because it's totally new and it has never been a part of my personality before" [P12].

Some couples talked with each other about changes they noticed:

> Well normally when I notice stuff, or my husband notices stuff, we talk it out….You know, he'll say, "I notice this and I notice that, what do you think?" [P06];

> I just asked her if she could pick up the dry cleaning or something, and she broke down….And then it hit me…it's not that she doesn't want to,…it's because it's too much for her….I think she thought it was HD-related, too. We kind of talked about it afterwards [C23].

However, other couples did not talk about HD: "We don't talk a lot about Huntington's or how we're feeling about it" [C17]. A few participants talked about the difficulty deciding whether changes are related to HD: "[I]t kinda gets kinda vague when you're at the presymptomatic point" [P06];

> [P]robably a lot of the things are subtle….I met her mom a little when her mom was alive…and some other HD people, and obviously those behaviors were precipitous…..[So] when something happens and it's just, you know, a very, very slow…degree, it's sometimes hard to notice it. Say, if you use an LED flashlight every day and as the batteries exhaust the light is getting dimmer and dimmer and you don't notice it until one day you say, "Wow, this is really dim" [C05].

Coping

Participants did not need to use coping strategies for changes that were not interfering with daily functioning. When changes did interfere with functioning, participants used several strategies to cope with them. Persons with prHD talked most often about active coping, information seeking, and using prescription medications. Companions talked most often about active coping, helping their partners, and acceptance. A more thorough examination of coping strategies using both qualitative and quantitative methods is presented in Chapter 4. The present analysis focuses on the link between changes, attributions and coping strategies.

All participants took action when they believed it would be helpful. Thus, there was a link between changes and the coping strategies participants used. Many participants used memory aids, for example, to cope with memory changes. Companions coped with partners' irritability by responding in ways that didn't escalate partners' irritation. Persons with prHD used prescription drugs to treat anxiety, depression, and sleep problems. Companions tended to help partners when partners became frustrated with cognitive tasks such as paying bills, planning, or doing new tasks. Both persons with prHD and companions said companions helped with daily tasks such as driving, cooking, housework, child care, and shopping.

Persons with prHD most often coped with social withdrawal by avoiding social interaction, although a few tried to socialize more. Companions coped with partners' social withdrawal either by encouraging them to socialize or attending social activities without them. Companions stated more often than persons with prHD that they coped by using acceptance. In fact, a person with prHD who did not attribute current changes to HD was ambivalent about accepting future changes: "I started taking vitamin supplements…just to ward it off….I'll try anything I guess,…yet at the same time I am realistic" [P04].

The link between attributions and coping strategies was not as evident. For the most part, participants coped directly with changes, regardless of attributions. For example, persons with prHD took medications to treat symptoms directly; they did not necessarily attribute them to HD. Persons with prHD relied on companions to help them, and companions helped partners with difficult tasks, regardless of whether either attributed changes to HD. Some persons with prHD and companions coped by making plans for the future; however, this coping strategy was not related to making attributions for current changes, but rather to anticipation of what they knew was coming. A few persons with prHD and companions used behavioral disengagement to cope with some

changes; however, they did so regardless of whether they attributed changes to HD or to other things.

For companions, there was a link between using acceptance or self-distractions as coping strategies and attributing changes to HD: “I don’t think there’s really much that can be done” [C17]; “[I]t doesn’t do me a whole lot of good to…push something that’s not going to get me anywhere” [C18]; “I know it’s coming. You know I know she has got the gene….I just take it one day at a time; that’s all we can do with it” [C19]. These responses indicate that because they attributed changes to HD, they could not be changed; therefore participants coped by accepting the changes.

One person with prHD stated she didn’t believe making attributions for changes was important in deciding how to cope with them:

> You know, you can’t definitely say it’s HD and you can’t definitely say it’s age….I guess my answer to that is, why does it matter what it is? The issue is, if you have symptoms, you have to deal with them [P12].

Evaluation and Reappraisal

In most cases, participants stated coping strategies were effective when changes were not severe. For example, using memory aids was effective most of the time. Both persons with prHD and companions stated that for the most part their attempts to cope with irritability were effective. Coping strategies were also effective regardless of what participants attributed changes to.

There were a few examples of cases when evaluations of coping strategies were related to attributions. For example, in one case, a companion gave his wife both visual and verbal reminders to purchase an item they needed at the store and his wife still forgot the item. The companion attributed his wife’s memory issues to HD because they were severe; his evaluation of the coping strategies as not effective was part of the reason he attributed her poor memory to HD.

When changes were more severe, coping strategies were less effective. Participants became frustrated, for example, when changes interfered with daily functioning. This was more evident in participants who attributed changes to HD, suggesting a link between attributions and evaluation of coping strategies: If strategies were less effective, participants might be more likely to attribute changes to HD. This phenomenon was apparent from both companions who talked about partners' excessive memory issues and forgetting shopping items.

Only two participants—a man with prHD and his wife—demonstrated reappraisal of an attribution based on the effectiveness of a coping strategy. After the husband started taking an antidepressant as part of a drug trial, he and his wife noticed positive changes which then led them to reappraise what changes were related to. When previously they thought changes were mostly related to aging or temperament, now they attributed many to HD:

> I found being social enormously difficult….I think that was one of the signs of Huntington's…because the medication seemed to end that [P02];
>
> [H]e was uncomfortable speaking in public, or getting more uncomfortable. He'd never been a comfortable public speaker, but it was stressing him out more….[Now he is] much less stressed out about having to make a public presentation….I assume [it's because of] the medication he's taking [C02].

It is notable that the participant did not select this coping strategy; rather it was selected for him as part of a drug trial to address symptoms in prHD. Although he was not told whether he received the placebo or the real drug, both he and his wife believed it was the real drug because it was effective. He continued to take the drug after the trial by asking his physician for a prescription.

Some participants demonstrated use of all the steps of the CSM—they noticed changes, made attributions, used and evaluated coping strategies, and demonstrated active processes in forming their representations related to noticed changes. Examples of

persons with prHD and companions using all the steps of the CSM process are presented in Table 5 and Table 6, respectively.

Discussion

Elements of the Common Sense Model

Participants noticed changes, made attributions, used coping strategies, and evaluated the effectiveness of coping strategies. Thus, all the elements of the CSM were apparent in the data. However, participants coped with changes when they interfered with functioning regardless of whether they made attributions for noticed changes. Coping strategies were usually effective unless changes were perceived to be severe. Some participants attributed changes to HD when active coping strategies were not effective, and coped by using acceptance or distraction. One couple reappraised an attribution based on the effectiveness of a coping strategy: When medication was effective in treating changes, the couple attributed changes to HD rather than to other things.

For the most part, changes were subtle and ambiguous. Participants formed attributions by comparing themselves to others with and without HD and to the former functioning of persons with prHD. The most common attributions for changes were aging, temperament or personality, and other stressors. Participants attributed changes to HD when they were more severe, there were more of them, and/or they were distinctive. These findings thus support the findings in a preliminary study by these researchers (Downing, Williams, & Paulsen, 2010).

Participants noticed several types of physical changes, including movement changes. It is surprising that participants did not always attribute movement changes to HD, even though HD has been traditionally diagnosed based on movement changes (Quarrell, 2008). Several participants also noticed changes in work function. This supports prior research that indicates work function is one of the earliest functional declines in persons with prHD (Paulsen, 2010).

Some notable differences in responses between persons with prHD and their companions were found. For example, persons with prHD were more likely to talk about problems in their relationships with their companions. One reason for this phenomenon might be fear of future dependence on their companions and concern whether companions will care for them when their disease progresses. Thus, they may be more anxious about their relationships. For example, more persons with amyotrophic lateral sclerosis expressed concern about dependency on their caregivers than their caregivers expressed about caring for them (Trail, Nelson, Van, Appel, & Lai, 2004). The use of instrumental and emotional support by over half of persons with prHD in the current study indicates that persons with prHD are aware of their need for support. Companions, however, expressed acceptance of changes they attributed to HD, perhaps suggesting they were willing to take care of their partners. In fact, companions gave many examples of how they helped their partners. Companions talked about planning for the future, indicating their intentions of providing for their partners. Companions who noticed changes that interfered with partners' functioning also acknowledged relationship issues; this suggests relationships may become more challenged as symptoms progress. In an older study on divorce and HD, for example, the majority of break-ups occurred in the first 2-3 years after HD diagnosis (Tyler, Harper, Davies, & Newcome, 1983). More recent data on divorce in HD was not found.

Another interesting finding is that not all couples were congruent in attributing changes to HD. While four couples were congruent in attributing changes to HD, three persons with prHD and three companions attributed changes to HD when their partners did not. This suggests that some individuals may prefer to attribute changes to HD before the changes are distinctive, while others prefer to wait until changes are less equivocal.

Evaluation of the CSM in Anticipated Illness: Prodromal Huntington Disease

The findings of this study indicate persons with prHD and companions have illness representations of diagnosed HD but not of the HD prodrome. While some participants used all the elements of the CSM, participants did not always make attributions for noticed changes or find it necessary to use coping strategies. For the most part, changes were not severe and participants could readily attribute them to things other than HD. This supports Leventhal and colleagues' assertion that people tend to delay making attributions or to make more benign attributions when changes are subtle and ambiguous (H. Leventhal et al., 1998).

Many of the dimensions of illness representation—consequences, duration, and controllability/control—were implicit in participants' attributions, active processes, coping strategies, and evaluations of coping, though they were not explicitly mentioned. Thus, illness representations may be better described as "in progress" in prHD, which may be relevant in other anticipated illnesses.

The first part of symmetry rule of the CSM was supported—participants sought to make attributions for noticed changes; however, they did not always do so. Several participants stated they didn't know what changes were related to. And one person was annoyed when asked what changes were related to, stating it didn't matter. The second part of the symmetry rule—that given a label, people will seek to find symptoms that match that label—was less supported. While a few participants talked about relating changes to HD based on knowledge that persons with prHD were going to develop HD, the majority of participants (32/46) did not attribute changes to HD. There may be various reasons for this apparent violation of the symmetry rule, including diminished insight in persons with prHD, use of denial or normalization as coping strategies, and lack of previous experience with prHD that inhibits formation of prHD illness representations.

Persons with prHD have demonstrated diminished insight into their own cognitive deficits, which has been associated with frontal brain dysfunction (Duff, et al., 2010a). On the other hand, diminished insight in persons with prHD does not explain why an equal number of companions also did not attribute changes to HD. It is possible that both persons with prHD and companions used denial as a way to cope with early changes in prHD. Participants may also try to normalize their experiences as much as possible. A more extensive discussion of denial and normalization as coping strategies will be discussed in Chapter 4.

Another explanation for why most participants did not attribute changes to HD is that they may have little knowledge of what prHD looks like. In a previous study (Downing et al., 2010), participants stated they did not expect changes because they were "presymptomatic," suggesting they couldn't have symptoms yet. In the current study, the researchers did not use the term presymptomatic to avoid inadvertently making this suggestion. The word "prodromal" was included in the consent documents in the title of the study, but in other places participants were told the study was for people who "tested positive for the HD gene expansion but were not yet diagnosed." Regardless, the majority of participants did not attribute changes to HD. This is particularly surprising given that all participants participate in HD-related research and are asked annually about changes.

The term "prodromal HD" is used by researchers, not by persons with the HD gene expansion. Persons with the HD gene expansion may not know they have any symptoms of HD until they are diagnosed with the distinctive motor signs. For them, HD may be more like an "on-off" switch rather than a disease with a long prodrome. While most persons with prHD grew up in families with an HD-affected parent, it is possible they did not associate early changes in their parents with HD. They may remember primarily the more florid symptoms that occurred later in the disease. Because fewer than a quarter of persons at risk for HD are estimated to undergo HD gene testing (Tibben, 2007), and the definitive test for the HD gene expansion was not available until 1993

(The Huntington's Disease Collaborative Research Group, 1993), it is possible most participants in the current study did not know their parents had HD until their parents were diagnosed based on motor symptoms. Moreover, seven of the 23 persons with prHD in the current study said they did not know their parent had HD until they themselves were adults. Stigma still prevents many persons from revealing their risk of HD, even to their own families (Wexler, 2010). Thus, participants might not have formed illness representations of prHD because they had no conscious past experiences with prHD.

Lack of conscious experience of prHD may partly explain why participants were often uncertain regarding how to make attributions for noticed changes. The uncertainty theory (Mishel, 1988) of illness states that persons are unable to make meaning out of changes if they do not recognize symptom patterns, if changes are not recognized as familiar to an illness, or if they are not congruent with their expectations or past experiences. Under these conditions, persons make their own appraisals of changes. This phenomenon may explain why in the current study participants attributed many of the same changes to different things. It is possible that the only consistent patterns persons with prHD recognize related to HD are the distinctive motor signs that constitute how HD has traditionally been diagnosed.

Overall, the CSM was a useful framework for exploring illness representations in prHD. Participants demonstrated use of the CSM elements, even though it is not clear from the data that they formed illness representations of prHD.

Implications

This study explored how persons with prHD and their companions experienced prHD. These findings indicate persons with prHD and companions were uncertain regarding what to expect prior to diagnosis. Participants had difficulty deciding what changes were related to and whether or not they were related to HD. The fact that many tried to get information from family members, written materials, researchers, and even

from the research materials themselves, suggests many participants would like more information regarding what changes to expect in prHD.

There has been an increasing trend for research participants to want to receive research results, raising ethical issues about potential benefits and risks of harm (Shalowitz & Miller, 2008). While a few participants in the present study tried to find out information about changes in prHD from researchers, there are no data regarding how many participants in the PREDICT-HD study would like to know whether they are exhibiting changes and whether these changes might be related to HD. However, these findings suggest that at least some participants would like more information. Persons who have undergone HD testing have stated they wanted to be tested in order to relieve uncertainty (Decruyenaere et al., 2003). However, other researchers have pointed out that testing positive for the HD gene expansion prior to motor diagnosis can be a way of trading one kind of uncertainty for another since persons are still faced with the uncertainty regarding when HD begins (Soltysiak, Gardiner, & Skirton, 2008). It may not be desirable to eradicate uncertainty because uncertainty itself may be a way for persons to maintain hope (Mishel, 1988). Indeed, persons who have decided not to undergo HD genetic testing have stated a desire to preserve hope as a reason to forgo testing (Quaid, Simes, Swenson, Harrison, Moskowitz, Stepanov, et al., 2008).

The authors of the CSM assert that illness representations are important because they influence the coping strategies people select. However, in the present study, most coping strategies were not related to attributions, suggesting it may not be important to know whether changes are related to HD in order to cope with them effectively. In the future, this could change. For example, by the time distinctive motor symptoms onset, 50% of neuronal death associated with HD has already occurred (Hannan, 2005). If treatments become available to delay or prevent HD symptoms, it might be necessary for persons with prHD to recognize HD affects them long before they have the motor signs in order for them to accept treatment, especially if the treatments have unpleasant side

effects, a phenomenon of the CSM known as the dose-dependent rule (H. Leventhal, et al., 1997).

The body of research related to changes in prHD is growing. The widening gap between what researchers know about prHD and what persons who have prHD know about themselves will inevitably force researchers to address ethical issues regarding duty to inform, risk of harm, and paternalism. Some have argued in favor of disclosing research results to participants (Fernandez, 2008). However, persons with prHD and their companions may also benefit from more general education on changes to expect in prHD.

Limitations

The findings from this study suggest that persons with prHD and their companions use elements of the CSM when trying to make meaning of changes. However, qualitative methods are unable to determine the relationships between attributions, coping, and outcomes. Statistical analyses using quantitative data, multiple regression, or path analysis could be used to facilitate these explorations. The addition of outcomes variables, such as measures of wellbeing, adaptation, relationship satisfaction, or quality of life, may be useful in these analyses to explore how attributions and coping strategies influence wellbeing.

Participants in this study may not be representative of most couples dealing with prHD. They all participate in ongoing HD-related research, indicating a possible bias toward a desire for HD-related information. Some persons contacted for possible participation said their companions would not participate in research; therefore, the couples who did participate may be more cohesive than other couples affected by HD. The low response rate to the mailed study invitations may also limit the representativeness of these data. Persons who did not answer their telephones or respond to mail may have been experiencing more severe changes such as apathy or social withdrawal that prevented them from responding. Thus, participants in the present study

may be experiencing fewer changes that interfere with functioning than persons who did not respond to recruitment requests.

It is also not clear if there are differences in participants' research experiences across PREDICT-HD study sites. For example, some participants stated they sought information and/or support from research staff during their study visits. The nature of this informal support may vary according to test sites and staff. In addition, not all participants may ask for help or advice and thus only those who ask may receive it. Finally, while we reached qualitative data saturation after 15 interviews, more participants may have added richness to the data.

Conclusions

The results of this explorative descriptive study indicate persons with prHD and their companions noticed functional changes in persons with prHD, made attributions for changes, coped with changes, and evaluated the effectiveness of coping. There was also limited evidence that persons with prHD and companions reevaluated illness representations based on the effectiveness of coping strategies. Although some elements of the CSM were only partially supported, such as the symmetry rule, these findings suggest the CSM is a useful framework for studying how persons with prHD and their companions make meaning of changes in the HD prodrome.

Figure 1. Recruitment Diagram

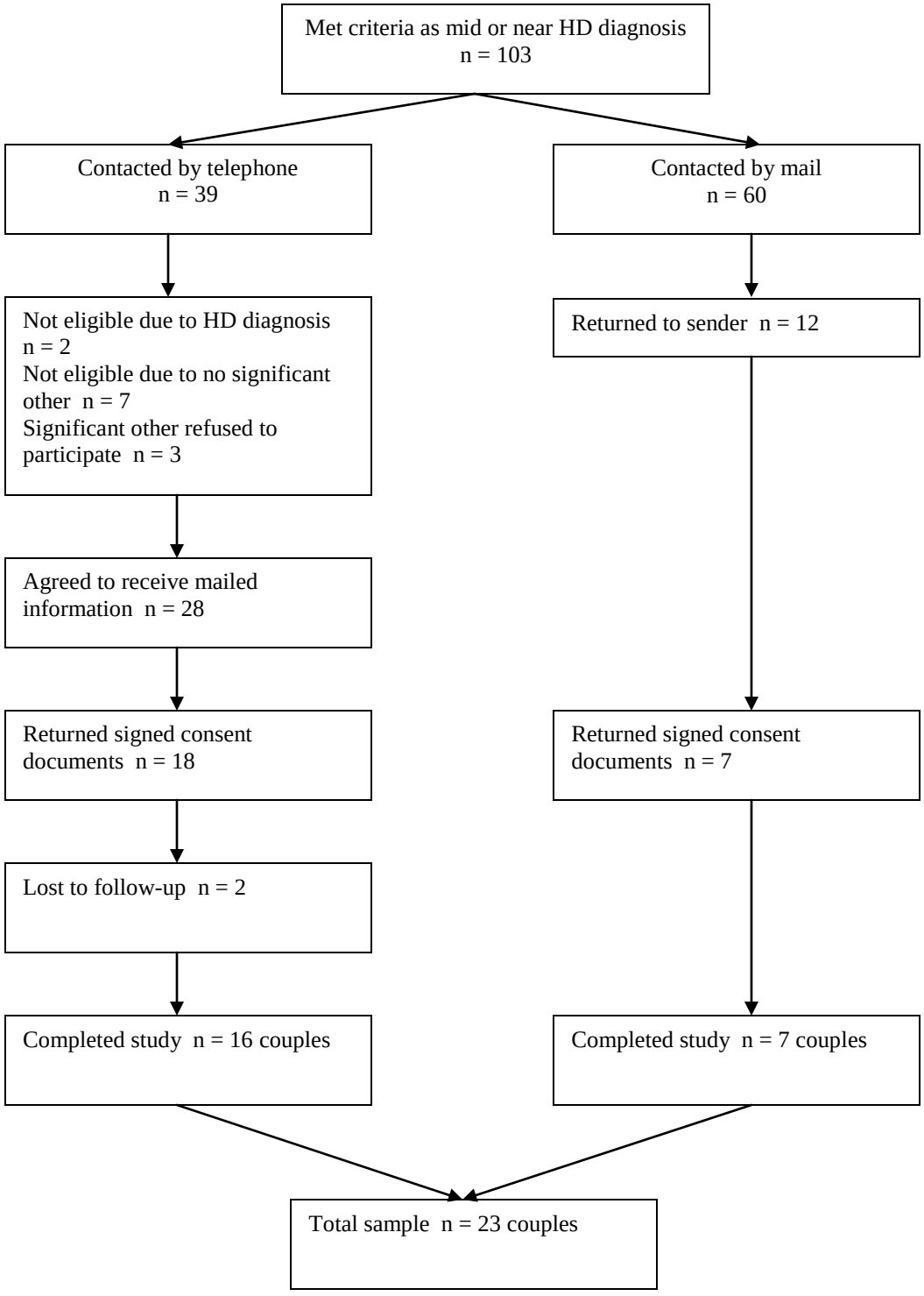

Table 2. Summative Content Analysis of Endorsed Changes

Changes	Number (%) who endorsed changes		Changes	Number (%) who endorsed changes	
	prHD	**Companion**		**prHD**	**Companion**
Memory	11 (48%)	11 (48%)	Fidgeting/ Involuntary Movements	3 (13%)	2 (9%)
Work	9 (39%)	7 (30%)	Slowing Down	3 (13%)	1 (4%)
Relationship	9 (39%)	4 (17%)	Miscellaneous Physical[c]	3 (13%)	1 (4%)
Mood	8 (35%)	9 (39%)	Home	2 (9%)	3 (13%)
Irritability/ Impatience	7 (30%)	8 (35%)	Clumsy/Dropping	2 (9%)	1 (4%)
Social life	6 (26%)	8 (35%)	Balance	2 (9%)	0
New Tasks	6 (26%)	6 (26%)	Spatial Judgment	2 (9%)	1 (4%)
Driving	5 (22%)	6 (26%)	Decreased Interest	1 (4%)	2 (9%)
Planning	5 (22%)	4 (17%)	Violent Behavior	1 (4%)	2 (9%)
Tired	5 (22%)	3 (13%)	Sleep	1 (4%)	2 (9%)
Miscellaneous Cognitive[a]	4 (17%)	1 (4%)	Obsessive Behavior	1 (4%)	3 (13%)
Ability to Focus	3(13%)	3 (13%)	Sex Life	0	2 (9%)
Miscellaneous [b]	3(13%)	2 (9%)	Gait Changes	0	2 (9%)
Multitasking	3(13%)	2 (9%)			

Each of these changes was mentioned by 1 participant:

[a]Persons with prHD: ability to do math , prioritizing, transitioning between tasks , paying bills; companions: Black and white thinking

[b]Persons with prHD: eating , hygiene, confidence , handwriting; companions: eating , self-absorption

cPersons with prHD: hand posture, fell out of bed, strength; companions: slowed speech

[d]Persons with prHD: grief, economy, hunger, long winters, HD discrimination; companions: poor judgment, distractions, culture, medication side effects

Table 3. Summative Content Analysis of Endorsed Attributions

Number (%) of Participants Who Endorsed Attributions		
Attributions	**prHD**	**Companion**
Aging	14 (61%)	7 (30%)
Don't Know	9 (39%)	5 (22%)
Personality	8 (35%)	8 (35%)
Huntington Disease	7 (30%)	7 (30%)
Stress/Busy	7 (30%)	5 (22%)
Relationship	6 (26%)	2 (9%)
Other[d]	5 (22%)	4 (17%)
Parenting	5 (22%)	3 (13%)
Depr./Anxiety	5 (22%)	3 (13%)
Hormones	4 (17%)	1 (4%)
Tired	3 (13%)	1 (4%)
Other Illness	3 (13%)	1 (4%)
Moving	2 (9%)	3 (13%)

Table 4. Links between Attributions and Changes

Attributions (In order of frequency of endorsement)	**Changes**
Aging	Memory, slowing down, feeling tired, difficulty multitasking, apathy, black/white thinking, decreased interest in sex, difficulty learning new things
Temperament/personality	Irritability, impatience, social withdrawal, work issues, memory
Don't know	Driving difficulties, memory, clumsiness, apathy, motor changes, social withdrawal, difficulty focusing, slowing down, obsessive behavior
Huntington Disease	Motor changes, anger, irritability, sleep problems, depression, clumsiness, social withdrawal, obsessive behavior, work issues, driving difficulties, eating issues, memory, apathy, difficulty learning new things, difficulty focusing, difficulty planning, difficulty multitasking
Stressors: moving, family, finances, job, being busy	Mood, irritability, housework, relationship issues, memory, work issues, tired, difficulty focusing, social withdrawal, difficulty multitasking
Other health conditions: menopause, depression, anxiety, other illnesses, injury	Mood, irritability, slowing down, motor changes, sleep changes, decreased interest in sex
Bad childhood with HD-affected parent	Depression

Table 5. Examples of CSM Process in Persons with prHD

Change	**Attribution**	**Active Process**	**Coping Procedure**	**Evaluation**
We fight quite a bit….I truly feel anger	I don't think it's age-related; it may be HD-related	I think maybe with this one issue it may be true anger in me, and that causes me to think [it's related to HD]…because people get older [but] they don't get more angry	[I]f we have too much conflict, we go see a…psychologist.	I think that works well
I just have been… experiencing depression, which I never really had problems with before	I don't think it's really 'cause of Huntington's….I think it's been more related to…problems with my mom	I guess I'm just not one of those people that walk into a room and…everybody goes "Oh,…she's fun and happy-go-lucky….[T]hat's never been pinpointed on me	I'll take a break and try to come back a little better….I'm on medications for it now	[T]here's those days where it's just…no matter what I do I can't…get out of it
I have just become more accident prone….I drop or spill	I don't know….I know it's triggered…when I'm tired	I've kinda looked stuff up and I don't know…[I]t gets …vague when you're at the asymptomatic point. If you point things to Huntington's or just to…tiredness….[T]here's times when I'm not tired and I'll grab a cup…[and] it just drops	I've pretty much accepted that it's going to happen…. I just…clean up the mess and do what I gotta do	I don't let it…stop my day or freak me out or anything
I have a hard time making connections with people	I've never been much of a real social person	I mean, I don't…go out to lunch with my girlfriend, I don't go shopping…I'd rather be with my kids….I'd rather be with my husband	I joined a women's group at church….I'm trying to go out of my comfort zone and do those things more now	I enjoy it

Table 5. Continued

Change	Attribution	Active Process	Coping Procedure	Evaluation
[At work] I do experience a bit of lethargy,…lack of focus,…trouble remembering details	I don't think it's [HD]….In talking to my psychiatrist, I don't think I'm worse than a typical 50-year-old that comes into an office….I think it's mainly age-related, but…the appropriate answer to that is maybe [it's HD-related]	You know, you can't definitely say it's HD and you can't definitely say it's age….I hope I never have to make that determination….[W]hen I'm clearly not performing, perhaps it will be obvious.	I'm starting to pay attention to my body and noticing things like being dehydrated and how that affects my lethargy and being able to focus... exercising versus not exercising,…eating sugar and all that kind of stuff	Absolutely [it is helpful]
Depression….Two summers ago I took a whole bottle of pills, hoping that would end my life	I think it's early onset Huntington's	My mother was depressed for a number of years and she also had Huntington's….She was not very nurturing…and when [my dad] was gone she physically abused us	I've been treated medically and therapeutically, through counseling…I'm walking 3 miles a day. And I do still smoke, and that's been my major coping mechanism	I've got a [medication] schedule now where for the first time I'd say for the last…2 or 3 weeks I feel almost my normal self again
[S]ometimes I think my double-tasking. I can't do as much as I used to	[E]verybody says, "Oh, it just comes with age. Don't blame it on Huntigton's." But…sometimes I do	I think, "Oh you know, this is probably the start of [HD]," but I don't know. People say they don't see anything in me	I just kind of don't let it bother me and I just keep going	[Effective]
I have noticed a difference…from the past year…losing interest a little bit quicker with things	I honestly don't know	I have never been an organized person…so that's…just part of being me….[and] dealing with stress of, yes, I'm going to turn into my mother one day…I think it's just a combination of it all	[L]aughter….[T]hat sounds just so cliché, but...you can make yourself miserable or you can just laugh about it and roll with the punches	how else are you going to deal with life?

Table 6. Examples of CSM Process in Companions

Change	**Attribution**	**Active Process**	**Coping Procedure**	**Evaluation**
Sometimes I felt like he just…overreacted to things	I was assuming it was just the early signs of the Huntington's	He's always been a little quick to fire…but it just seemed to be stronger	[A]s soon as he kind of had that strong reaction, I just kind of…back off and sort of almost under react	Sometimes it worked well and sometimes [he] took it as I wasn't really caring
I had tickets to [a concert] last weekend, and he didn't want to go	I don't know if it's his personality or if it's a change	I'm getting more I think "no"s. He's always known what he's wanted….I just feel like those are gonna be the personality traits that I think are gonna get just even more and more sharper	I paid to go and so I went…with my sister	I always reach the point where it's okay that he says no
[S]ometimes her outbursts are very, very personal. She gets really caustic. And sometimes she's violent	I think they're related to her…Huntington's	I've got documentation that…tells 10 easy ways to identify Huntington's symptoms and she's got many of those. For example, her outbursts…they're identical to what the description would be	[T]he one thing that I've learned to do is to not respond to them….[I] just say to myself, "She's really sick…and that's where this is coming from"	I'll tell you something, it's really, really hard on me
[L]ittle movements… maybe a little jerkier, a little wider…walking is a little less smooth	I think these are Huntington's changes… just the early stages	I remember [his] father and his sister, and …I don't think these are just… normal…aging changes. I think they're the Huntington's chorea….I think they'd be noticeable to somebody who didn't know about the Huntington's	I don't think there's really much that can be done. I mean it's not to the point where he'd need medication to control them	I just look at it and say this is, you know, perhaps a sign that things are gonna start happening

Table 6. Continued

Change	Attribution	Active Process	Coping Procedure	Evaluation
[T]here's been slow changes….She's more clumsy, she has less interest in doing things, …lack of motivation, forgetfulness	I think it's the Huntington's	[H]er and I are both educated into the whole Huntington's thing, and know what to look for.	It doesn't do me a whole lot of good to…push something that's not going to get me anywhere….We just try to help her out	I get frustrated
She's more easy to get irritated	I know that's part of the symptoms	[Y]ou know that [HD newsletter] that comes out? Several people talk about how…the husbands or the wives they get a little more irritable as…it progresses	I try to keep my conversations short with her 'cause all she wants to do is argue	Sometimes [it works]
Worsening depression	I'd say life stresses…[or] the fact that she is gene positive for Huntington's	[W]hat role does depression play with Huntington's, or Huntington's play with depression? …I just don't know…if her depression is the early manifestation or just a side effect of having the gene	[W]ork, and I have hobbies—reading, reading the paper,...I play golf. You know, mainly things like that	You don't cope with those things…you do the best you can, you know?
Getting frustrated learning new things	Huntington's	[O]nce you find out that you have Huntington's in your family, sometimes you might start looking for things and blaming it that way	[F]or the most part I will attempt to get her to do something new	[S]ometimes she willingly jumps in and tries it and sometimes she just [gets] frustrated
[W]e're getting ready to go down South and visit some friends…and I know right now that she will fret more than what I would consider normal	I think that's probably Huntington's related	I guess it's just something that's slowly developed over time, and I'm making that assumption. I can't tell you that I have a concrete thing that says "aha!"	Get out of the way!	It's better than if I were to try to help. I'd probably get my head cut off

CHAPTER 4

COUPLES' COPING IN PRODROMAL HUNTINGTON DISEASE

Abstract

Huntington disease (HD) is a progressive neurological disease with typical onset in middle adulthood. There is a long prodromal phase (prHD) in which changes in behavior, cognition, and motor function begin to occur up to 15 years prior to diagnosis. Changes are subtle at first and may be difficult to attribute to HD. Little is known regarding how persons with prHD and their companions cope with changes in prHD. This study uses quantitative and qualitative methods to explore and describe how persons with prHD and their companions cope with noticed changes. Twenty-three couples were interviewed using a semi-structured interview guide and the Brief COPE scale. Both interview and Brief COPE data showed persons with prHD used more coping strategies than companions. On the Brief COPE, persons with prHD used acceptance, emotional support, and planning most frequently; companions used acceptance, planning, and active coping. The least frequently used coping strategies by both persons with prHD and companions were denial, behavioral disengagement, and substance use. Three major themes from the qualitative interview were identified: trying to fix it, can't fix it, and not broken yet. Qualitative interviews revealed some coping strategies that the Brief COPE did not measure: Persons with prHD used prescription medications, dyadic coping, hope, and self-monitoring; companions used hope and helped their partners with prHD. Many of the coping procedures were effective, especially when changes were not severe. Participants who noticed more severe changes talked about acceptance and distraction as coping strategies. Persons with prHD and their companions may benefit from learning about coping strategies participants stated were helpful. Persons with prHD may benefit from using prescription medications to treat mild depression, anxiety, and sleep

disruption. Couples may benefit from counseling that helps them use coping strategies that are helpful when changes can be fixed and to accept things they cannot fix.

Introduction

Huntington disease (HD) is a progressive neurological disease characterized by behavioral, cognitive, and motor function and premature death. It is caused by an expanded trinucleotide (CAG) repeat on chromosome 4 and has a dominant inheritance pattern—people with an affected parent have a 50% chance of developing the disease (Walker, 2007). Average age of diagnosis is between 35 and 55 (Quarrell, 2008) and is based on the presence of distinctive motor signs—chorea, dystonia, and impaired voluntary movements (Hogarth, 2003). However, researchers are able to detect changes in brain structure, cognition, and motor function in persons who are positive for the HD gene expansions up to 15 years before diagnosis (Paulsen, 2010). Little is known regarding how people who have tested positive for the HD gene expansion and their companions cope with functional changes that may impact their daily lives before diagnosis, a period that has been referred to as prodromal HD (prHD) (Paulsen, 2010).

Persons with prHD and their companions do not necessarily recognize subtle functional changes or if they do, they may not attribute them to HD (Downing et al., 2010). Family members have noticed changes in persons with prHD, including irritability, impaired judgment, difficulty sleeping, and problems with balance (Williams et al., 2007), but expressed uncertainty regarding whether changes were related to HD; they also stated a desire for more information regarding what to expect prior to diagnosis. Even less is known regarding how persons with prHD and their family members cope with changes. The purpose of this study is to use qualitative and quantitative methods to describe coping in persons with prHD and their companions. The research questions include: 1) How do persons with prHD and their companions cope with perceived functional changes in prHD? 2) Do persons with prHD and companions use similar or

different coping strategies? 3) Are the coping procedures effective? 4) Are coping strategies different for those who attribute changes to HD? These data may provide information regarding coping procedures that may be used to develop interventions to improve wellbeing for persons with prHD and their companions.

Theoretical Framework

Coping is considered important to quality of life because the ways people cope impact their mental and physical health (Lazarus & Folkman, 1984). Coping is a component of Leventhal and colleagues' Common Sense Model of Illness Representation (CSM; H. Leventhal et al., 1998), a framework developed to explore the process of how people make sense of health-relevant information, select coping procedures and evaluate their effectiveness. The model derives its names from the "common-sensical" connection between illness representations and the selection of coping procedures (H. Leventhal et al., 1998, p. 722). Leventhal and colleagues (1998) define health-related coping as "the cognitive and behavioral actions we take (or do not take) to enhance health and to prevent, treat…and rehabilitate from illness" (p. 722). People select coping procedures based on "If-Then" rules (e.g. "IF my illness is caused by stress, THEN I will work to reduce my stress level").

There have been many efforts to characterize types of coping and coping styles. Many are based on the work of Richard Lazarus who defined coping as the "constantly changing cognitive and behavioral efforts to manage specific external and/or internal demands that are appraised as taxing or exceeding the resources of the person" (Lazarus, 1999, p. 110). Lazarus and Folkman (1984) distinguished two major types of coping: emotion-focused and problem-focused. Emotion-focused coping relates to the attempt to relieve distress through emotional responses such as avoiding thinking about the stressor, or minimization of the threat. A stressor is a "situation that is appraised by the person as taxing or exceeding his or her resources and endangering his or her well-being" (p. 19).

Generally, emotion-focused coping does not change a stressor; it just reacts to the stressor. Problem-focused coping, on the other hand, is focused on changing the stressor through action. These include cognitive processes such as defining the problem, weighing alternatives, and planning as well as taking action. In HD caregivers, "passive" coping strategies have been associated with greater distress than active coping strategies (Decruyenaere et al., 2005). Some have suggested that emotion-focused coping items on standard measures of coping are biased toward emotions that are maladaptive and they suggest a new scale that incorporates adaptive types of emotional coping, including acknowledgement, processing, and expression (Stanton, 2000).

Measurement of Coping

Quantitative measures of coping allow for systematic assessment of coping and facilitate comparison of coping strategies within and across samples (Schwartzer & Schwartzer, 1996). Several tools have been developed to measure coping, many based on Lazarus and Folkman's stress and coping theory. Two frequently used scales include the Ways of Coping Questionnaire (WCQ; Folkman & Lazarus, 1988) and the Coping Orientations to Problems Experienced (COPE; Carver, Scheier, & Weintraub, 1989). However, a limitation of both these questionnaires is length. Carver (1997) subsequently created the Brief COPE to address subject burden. The Brief COPE consists of 14 2-item scales which represent different coping methods. While Carver states the scales are distinct, other researchers attempt to group the scales into larger categories of coping styles to facilitate analyses, including "adaptive" and "maladaptive" (Meyer, 2001), and problem-focused, emotion-focused, and dysfunctional coping (Cooper, Katona, Orrell, & Livingston, 2006), although Carver discourages this (Carver, 2007).

Dyadic Coping

As many authors have recognized, coping with illness is rarely a solitary process. When one partner in a couple is affected by illness, the other partner is also affected

(Kenny & Cook, 1999). Each member of the couple may cope in unique ways, and they also respond to each other's coping. Coping in dyads has been characterized in terms of congruence and complementarity (Badr, 2004). Congruence refers to members of a dyad using similar coping strategies. In some instances, congruence has been associated with more favorable outcomes. For example, when persons with multiple sclerosis and their spouses both used problem-focused coping, this was associated with lower levels of distress and depression in both spouses (Pakenham, 1998). Women with breast cancer reported less distress when their spouses used levels of emotion-focused coping similar to their own (Ben-Zur, Gilbar, & Lev, 2001).

Complementarity, on the other hand, recognizes that it is not necessary for both members of a couple to cope in the same way in order for dyadic coping to be effective. For example, in the breast cancer study, incongruence in the use of problem-focused coping strategies did not cause distress in either member of the couple (Ben-Zur et al., 2001). Couples showed better dyadic adjustment when they exhibited opposite levels of protective buffering and avoidance coping (Badr, 2004). In a qualitative study, couples who characterized coping as a dyadic process, regardless of whether either or both members used emotion-focused or problem-focused coping strategies, identified positive benefits during a woman's breast cancer experience, including increased closeness; couples who characterized coping as individual processes typified by avoidance strategies were less able to identify positive benefits (Kayser, Watson, & Andrade, 2007). Thus, both partners do not have to cope in the same way in a successfully coping dyad; however, conceptualizing a stressor as a joint stressor may result in more successful outcomes. In fact, psychological symptoms were higher in couples when both used the emotion-focused strategy of escape-avoidance while coping with parenting adolescents (Giunta & Compas, 1993). Wives' use of escape-avoidance was associated with high psychological symptoms in both parents, but husbands' use of escape-avoidance only predicted their own distress.

This last finding suggests the possibility of gender differences in coping. Women may use more emotion-focused coping and men may use more problem-focused coping (Tamres, Janicki , & Helgeson, 2002). Other gender differences have been found. For example, when one member of a couple had cancer, women reported higher psychological distress and lower quality of life regardless of whether they were the patients or the caregivers; only men who were patients scored high on psychological distress and low on quality of life (Hagedoorn, Buunk, Kuijer, Wobbes, & Sanderman, 2000). Couples' coping strategies in illness may depend on whether the person with the health condition is male or female (Badr, 2004): Men who were ill used more problem-focused strategies; however, when their wives were ill, they engaged in emotion-focused strategies mostly designed to protect their wives them from negative thoughts and experiences. In couples who have had an unsuccessful attempt at infertility treatment, husbands' problem-focused coping was associated with less distress in wives before fertility treatment; after unsuccessful treatment, women who were low on emotion-focused coping before treatment had less distress if husbands used emotion-focused coping (Berghuis & Stanton, 2002). The authors state these findings suggest emotion-focused coping may be more helpful when stressors are uncontrollable. These studies emphasize the complexity of dyadic coping, including the finding that different coping styles may be effective at different points during a stressful situation.

Couples Coping with HD

There have been several studies looking at the impact of HD predictive testing on couples. For most couples, receiving a positive genetic test did not adversely affect their relationship beyond the initial shock of the results (Richards, 2004); companions in this study coped by seeking information and monitoring their partners for symptoms. Some persons with prHD minimized the impact HD would have on couples' lives, while companions and other persons with prHD stated HD would significantly impact their

lives. Dyadic adjustment after HD predictive testing did not decrease at 5-6 months and 18 months after testing for couples in which the at-risk partner tested positive, suggesting that, at least in the early period after testing, couples were still coping well (Richards & Williams, 2004). It is possible that couples who go through with testing and test positive have more positive relationships initially than couples who decide not to undergo genetic testing (Quaid & Wesson, 1995). Five years following HD testing, persons who tested positive reported greater dyadic consensus and satisfaction than their partners (Decruyenaere et al., 2004). Less in known about how persons with prHD and their companions cope with everyday functional changes, although some partners of persons with prHD have stated it was difficult to cope because they did not know what to expect regarding early symptoms (Evers-Kiebooms, Swerts, & Van Den Berghe, 1990)

Helder and colleagues (2002b) explored coping in 90 spouses (54.4% female) of persons with diagnosed HD. The most commonly used coping strategies by spouses using the COPE scale were acceptance, positive reinterpretation and growth, active coping, and planning. In a qualitative exploration of coping in ten HD caregivers (60% female), avoidance coping was common (Lowit & van Teijlingen, 2005). These findings raise two important points: First, not all uncontrollable situations elicit the same coping response, and generalizations across illness situations may not be possible; second, quantitative and qualitative methods may reveal different types of coping strategies even within the same illness.

Qualitative Measures of Coping

Some researchers believe questionnaires are inherently inadequate to asses coping; instead they recommend semi- structured interviews (Coyne & Gottlieb, 1996). Quantitative measures may not adequately capture dyadic coping because members of couples cope both as individuals as well as part of a couple, a distinction coping scales cannot capture (Badr, 2004). Quantitative measures also limit the number and types of

coping procedures that can be measured, while interviews can reveal strategies unique to the stressor of interest. For example, family caregivers of people with HD coped by anticipating the death of the HD-affected relative, and using prescription medications (Williams et al., 2009).

Since coping is an active process (i.e. based on "If-Then" contingencies), qualitative methods are more suited to capturing this dynamic process (Marteau & Weinman, 2006). Furthermore, qualitative methods allow participants to evaluate their coping procedures, which is another aim of this study. Finally, coping is contextual (Lazarus, 2000); thus, quantitative measures may not be able to account for context. Not all situations are alike and rarely do individuals have only one stressor. This may lead researchers to make false assumptions when interpreting quantitative coping measures.

Methods

Design

A mixed methods strategy was chosen in order to take advantage of both quantitative and qualitative methodology to explore coping in persons with prHD and their companions. The theoretical thrust of the study purpose is inductive; thus the core component of this study was qualitative (Morse & Niehaus, 2009), while the quantitative component facilitated systematic assessment of coping and comparison of strategies between persons with prHD and companions.

Sample

Potential participants were identified in collaboration with a PREDICT-HD study coordinator (Anne Leserman). PREDICT-HD is a multi-site longitudinal study designed to identify and track markers of HD during the prodromal period in persons who have independently undergone HD genetic testing (Paulsen, 2010). Purposeful criterion sampling focused on identifying participants with criteria of interest who were likely to

provide rich information using qualitative methodology (Patton, 2002). Inclusion criteria included: persons with prHD estimated to be ≤15 years from HD diagnosis using an algorithm based on CAG length and current age (Langbehn et al., 2004), ≥21 years old, and English-speaking; each person with prHD had to have a spouse or significant other who was also willing to participate. Couples were excluded if the companion had a severe chronic health condition. A complete description of the PREDICT-HD study and the sampling method for the current study was described in Chapter 3. A recruitment diagram is presented in Chapter 3, figure 1.

Procedure

The study was approved by the University of Iowa Institutional Review Board. Prospective participants were contacted by telephone or by mail. Twenty-three couples agreed to participate in this study. Prior qualitative studies using semi-structured interviews to explore illness representation in other illness using the CSM included sample sizes of nine (Harman & Clare, 2006) and 30 (Walter & Emery, 2006); researchers in both studies stated they reached qualitative data saturation. In the current study, data saturation was reached after interviewing 15 couples. However, all interested couples were interviewed in order to add richness to the qualitative data and facilitate meaningful interpretation of quantitative data.

Measures

Demographic Information

Demographic information was collected prior to beginning the semi-structured interviews and included age, gender, whether participants with prHD had children, and how long companions had known their partners.

Semi-structured Interview Guide

The semi-structured interview guide consisted of open-ended questions regarding how the person with prHD was functioning, whether participants noticed any changes in any of the following areas of function: work, home, relationships, social life, physical activities, driving, planning, memory, planning, and performing new tasks. Items came from functional changes that persons with prHD and family members have previously endorsed (Downing et al., 2010; Williams et al., 2007) and changes identified by researchers in a clinical setting (Paulsen, 2010). The interview guide also included questions regarding to what participants attributed any changes. In the CSM, attributions guide the coping procedures people select. Participants were also asked whether they considered their coping strategies to be effective. See Appendix A and B for complete interview guides.

The Brief COPE

Following the semi-structured interview, participants responded orally to the Brief COPE (Carver, 1997). The Brief COPE consists of 28 questions—two questions for each of 14 scales: active coping, planning, positive reframing, acceptance, humor, religion, using emotional support, using instrumental support, self-distraction, denial, venting, substance use, behavioral disengagement, and self-blame. Each item on the Brief COPE is comprised of an "I" statement regarding how frequently participants used each coping strategy. Because the researcher orally administered the Brief COPE, "I statements" were changed to "you statements:" Response choices ranged from "You usually don't do this at all" to "You usually do this a lot," and numerically rated from 1-4. Internal reliability for the Brief COPE scales provided by the author ranged from .57 (acceptance) to .90 (substance use) (Carver, 1997). The entire measure is presented in Appendix C. The Brief COPE has been used by researchers to assess coping in a variety of illnesses, including

perinatal depression (de Tychey et al., 2005) cystic fibrosis (Wong & Heriot, 2008) and mental illness (Meyer, 2001).

Ostensibly, the Brief COPE can be used to assess either dispositional coping style or situational coping style; the author of the scale permits researchers to adapt the instructions to suit their own research purposes (Carver, 2007). In the current study, the measure was used to assess dispositional coping because it was anticipated that some participants would not have noticed any changes at the time of the interview; therefore, participants were thus asked how they typically dealt with stress in the past month. Carver (2007) states the scale is not intended to identify coping styles, per se. Nor should it be used to identify emotion-focused and problem-focused coping. Rather, he suggests researchers analyze each scale separately and compare them with variables of interest.

Data Management

Consent documents and hardcopy interview notes were kept in a locked filing cabinet separate from interview notes. Interview guides and transcripts were identified by participant number only. Interviews were audio-recorded using a digital audio recorder and downloaded to the researcher's password-protected laptop. Interviews were then deleted from the audio recorder. An IRB-approved transcriptionist transcribed the interviews verbatim and the researcher verified them for accuracy. All identifying information was replaced with generic terms such as "name," "place," etc.

Transcripts were imported into NVivo8 (QSR, 2000) for qualitative data management. Qualitative data were managed using strategies outlined by Knafl and Webster (1988). Descriptive coding categories were developed using the interview guide and the Brief COPE as a framework. For example, "changes," "attributions," "active process," "coping strategy," and "evaluation" and Brief COPE scales "acceptance," "planning," "humor," etc. were descriptive coding categories. A codebook was developed

and included the definition of coding terms along with criteria for each coding category, and a sample excerpt for each.

Quantitative data were managed using Excel spreadsheets and PASW Statistics Developer (SPSS, 2009), a statistical software package. All programs were managed on password-protected computers.

Data Analyses

Quantitative Data Analysis

Demographic and Brief COPE data were entered into PASW to create descriptive data tables with ranges, means, and standard deviations. The Brief COPE was not intended to generate a composite coping score (Carver, 2007). However, higher scores on the Brief COPE indicate more frequent use of coping strategies, which conceptually could be an indicator of greater perception of demands using Lazarus' definition of coping. Therefore, mean total scores on the Brief COPE were calculated to indicate how frequently participants used coping procedures. Mean coping for each of the 14 2-item coping strategies were averaged to retain the 1-4 range.

A dependent *t*-test was used to compare differences in frequency of coping and types of coping strategies used between participants with prHD and companions. An independent *t*-test was used to compare frequency of coping between male and female participants to test for gender differences. Another independent *t*-test was used to test for differences in frequency of coping and whether participants attributed changes to HD. Levene's tests for equal variances indicated variances were not significantly different between persons with prHD and companions, between female and male participants, and persons who made HD attributions and those who did not, making parametric *t*-tests appropriate. Effect sizes for differences found in the *t*-tests were calculated using Cohen's *d*. Pearson correlation coefficients were used to test for congruence of coping

strategies between participants with prHD and companions and to test for correlation between frequency of coping and number of HD-related changes.

Qualitative Data Analysis

Data were analyzed using descriptive interpretive methodology (Thorne, Kirkham, & MacDonald-Emes, 1997). The purpose of descriptive analysis is "to sensitize the reader to the viewpoint of a particular group," (Knafl & Howard, 1984, p. 20), which is important when presenting a topic about which little is known (Knafl & Webster, 1988). Interpretive analysis takes into account the active role of the researcher in creating meaning out of descriptive data (Lowenberg, 1993). Transcripts were first coded by two of the researchers (ND and JW) who identified coping strategies using the Brief COPE as a coding frame; from those descriptive categories the researchers identified major themes (Knafl & Webster, 1988). They discussed descriptive and interpretive codes until they reached 100% agreement to establish descriptive validity (the descriptive codes were apparent in the data to both researchers) and interpretive validity (the interpretations of the data made sense to both researchers) (Sandelowski, 2000).

Mixed Methods Data Analysis

Summative content analysis (Hsieh & Shannon, 2005) of the qualitative data facilitated comparison with quantitative results, including the number of coping strategies described by participants and comparison of strategies with the 14 coping strategies from the Brief COPE. Two researchers (ND and JW) independently coded the transcripts from the first ten couples interviewed using the coping strategies from the Brief COPE as a coding frame, and adding new codes when a coping strategy did not seem to fit into one of the Brief COPE strategies. Definitions of each strategy were taken from the items in the 14 Brief COPE scales. For example, active coping was defined as "concentrating efforts on doing something" and "taking action" (Carver, 2007). The researchers discussed their coding until 100% agreement was reached.

The purpose of the content analysis was to compare coping strategies between data from the Brief COPE with data from the qualitative interviews for both persons with prHD and companions. This method provided insight into the ability of the Brief COPE to comprehensively capture coping strategies in prHD. Participants who indicated on the Brief COPE that they used the strategy at all (a score of 2 or higher) were counted as having used that strategy.

Results

Demographic Data

The majority of the 23 participants with prHD were female (N=17; 73.9%); companions were mostly male (N=16; 69.6%). Most couples were married (N=21); two were committed partners. Mean age of participants was 48.96 (SD=11.80; range 33-78); mean age of companions was 49.96 (SD=10.47; range 31-67). Couples had known each other a mean of 22.46 years (SD=13.32; range=0.5-47 years; median=21 years). Only two participants with prHD did not have children. Therefore, comparisons between participants with children and those without were not possible.

The Brief COPE

The three most frequently used coping strategies by all participants were acceptance (M=3.23; SD=0.14), planning (M=2.75; SD=0.19), and emotional support (M=2.68; SD=0.16). The means and standard deviations for the three most frequently used and least frequently used coping strategies by participants with prHD and companions are presented in Table 7. The three most frequently used coping strategies by persons with prHD were: acceptance, emotional support, and planning; the three least frequently coping strategies were: substance use, denial, and behavioral disengagement. For companions, the three most frequently used coping strategies were: acceptance, planning, and active coping; the three least frequently used coping strategies were:

behavioral disengagement, denial, and substance use. The number and percentage of participants who endorsed using each strategy at least "a little bit" (a score of 2 or higher) is listed in Table 8.

When the frequency of use of coping strategies was totaled for the entire measure and transformed to a 1-4 scale, persons with prHD had a mean total score on the Brief COPE of 2.26 (SD=0.43); companions had a mean total score of 1.91 (SD=0.44). Dependent *t*-tests showed participants with prHD used coping strategies more frequently than companions (t=3.11; p<.01; d=.81). An independent *t*-test for gender differences revealed that females used coping strategies more frequently than males (t=3.21; p<.01; d=0.76). Therefore, differences in frequency of using coping strategies in this sample may be more related to gender differences than to being either a person with prHD or a companion. An independent *t*-test to determine whether frequency of coping was related to making HD attributions was not significant (p=0.12).

Significant differences in frequency of coping strategies used by persons with prHD and companions included: self-distraction (t=2.79; p=0.01; d=0.84); emotional support (t=4.24; p=.00; d=1.28); instrumental support (t=2.26; p=0.03; d=0.68); and self-blame (t=2.21; p=0.04; d=0.68). In all cases, persons with prHD used these coping strategies more frequently than companions. However, while more persons with prHD used self-blame and self-distraction, they used them infrequently. Couples were congruent in their use of three coping strategies: active coping (r=0.46; p<0.03); planning (r=0.45; p<0.03); and religion (r=0.51; p<0.02). The correlation between number of HD attributions for changes and frequency of coping strategies was not significant.

Qualitative Results

Interviews lasted an average of 29 minutes for persons with prHD (range 9-58 minutes) and 25 minutes for companions (range 13-48). Length of interviews was related in part to the number of changes participants noticed and how talkative participants were.

In the descriptive analysis, seven persons with prHD and seven companions attributed changes to HD. Most couples (N=13) were congruent in their belief that there were no HD-related changes. Four couples were congruent in their belief that there were HD-related changes. Three persons with prHD and three companions attributed changes to HD when their partners did not. Participants attributed changes most often to age, temperament, and other stressors.

While most participants did not attribute changes to HD, most did notice changes. The most common changes mentioned by both persons with prHD and companions were physical changes (tired, slowing down, fidgeting, and dropping things), short-term memory problems, mood changes and irritability, work stress and employment changes, cognitive changes (difficulty focusing and finishing tasks, apathy, and difficulty with math), relationship issues, problems driving, decreased socializing, and difficulty planning and doing new tasks. The number of participants who endorsed each of the coping strategies identified in the qualitative interviews is shown in Table 8. Greater details regarding noticed changes and attributions for changes were presented in Chapter 3. In the interpretive analysis, the researchers identified three major themes related to coping with changes: *trying to fix it*, *can't fix it*, and *not broken yet*.

Trying to Fix It

The most common theme was trying to fix changes that interfered with functioning. Persons with prHD described actions they took to cope with changes, including: using memory aids; taking prescription medications; working on their relationships; seeking advice, assistance or information; making plans for the future; and self-monitoring: "I've been trying to find some type of work that works for my brain that I can still do" [P15]; "I have a hard time making connections with people….I'm trying to go out of my comfort zone and do those things more now" [P06];

> I make more lists now than I used to….[If] I put something on the stove in the kitchen, I don't leave the kitchen. I have burned

> up three tea kettles by leaving the kitchen and going and doing something else. [If] I do put something on the stove [I] have to actually sit in the kitchen [P01];
>
> I'm very heavily medicated….I take the Effexor and Trazodone for depression. I take Sonesta to help me sleep. I take ADD meds also….I've been doing a lot of therapy, working with a psychiatrist….You know my goal is to fight it and keep my mind as active and healthy as I possibly can [P12];
>
> I'm one of those people that don't drive with the radio on….I pay very [good] attention to all the things because I don't drive a lot so I don't take it for granted [P04];
>
> [A] lot of times I've asked questions to the other people in the other studies, you know, like…"Is this normal?"...[A]nd [I] look stuff up [P06];
>
> [M]y wife knowing that I tested positive, and all my daughters knowing I tested positive, it's open to us. And so…the communication has been real, you know, forward with, you know, if I become a problem to deal with then they have brought it to my attention so that I can try to, you know, help everybody cope better [P07];
>
> I've been kind of tired on and off lately.... [W]hen I get tired I have a tendency to get a little cranky….I try to take a nap when I can…or take a little time off of work here and there [P09];

Companions described actions they took to cope with changes, including responding to partners' irritability, helping their partners, planning for the future, and seeking information and advice: "I push her as much as I can to exercise because I know that that's gonna be critical when she does get onset" [C05]; "I pay all the bills because she's not good at that anymore" [C18];

> What I want to do is leave her the same income we have right now, and…I don't want to eat away at investments and things like that, and then her come up short when she is trying to live off of [them] [C04];
>
> [L]iving with someone for so long, you sort of get a sense of what they are going to react to, and…as soon as he kind of had that strong reaction, I'd just sort of…back off and sort of under react [C02];
>
> [She's been] dropping things at home, tripping, you know, walking….[W]e just try to help her out. And it's like, you know, "Let me carry that," you know? [C18].

For participants who took action to fix changes, their coping strategies were effective. Strategies worked well for changes that were not severe and were manageable with simple interventions such as memory aids for persons with prHD. Companions were able to manage partners' irritability by carefully responding when partners were irritable. Some companions also took action to plan for partners' future disability.

Can't Fix It

Some participants did not try to fix things. These coping strategies fell into two major categories: Some persons had tried to fix things and had given up when efforts weren't effective; other persons accepted right away there were some things that couldn't be fixed. Some persons with prHD coped by giving up or avoiding situations that were problematic, using distractions or accepting things they couldn't fix: "I just pray, and I try to put it out of my mind and go on" [P04];"I pretty much hate [doing new things]…. You know, if it's something new, I'll avoid it" [P15];

> Honestly, I don't know. It's like a lack of confidence in certain things, and not an actual lack of confidence in my ability….[S]ome things I don't feel like I can do anymore [P04];

> I have just become more accident prone, you know, drop stuff…like if I'm stressed out or tired….So I mean, I know that it's just something that's going to start affecting my everyday life. I'm not like spooked by it or anything. I've pretty much accepted that it's going to happen [P06];

> We do make jokes about it—the whole Huntington's thing. Or, you know, being clumsy or whatever. Because that's just, how else are you going to deal with it? [P18].

Companions also coped with stressors they couldn't fix by giving up, avoiding stressors, using distractions, or accepting: "I've learned the things *not* to do….[I] definitely don't try to fix it" [C12]; "Our sex life sucks….I get depressed; I experience being depressed a lot. I mean that's my way of coping" [C15];

> I had tickets to [a concert] last weekend and he didn't want to go….I'm getting more, I think, of the 'no's'….I paid to go, and so I went, you know, with my friend….I just ignore it. I'm finding

> the last couple of years…if he says no, I just go on with my life. I mean, he's not stopping me [C07];
>
> [E]ssentially coping is taking your mind off of [things], looking at things on the internet, reading,…just whatever it is, not to mention trying to help your spouse,…doing what you can [C20];
>
> I've noticed…little… movements…maybe a little bit jerkier, a little wider….I don't think there's really much that can be done. I mean it's not to the point where he'd need medication to control them….I just look at it and say, this is, you know, perhaps a sign that things are gonna start happening [C17];
>
> The only thing I notice anymore is anytime you talk to her, all you do is end up arguing with her all the time, you know?...I usually try to stay away when she gets in her moods, so I stay away from her [C19].

Participants who acknowledged there were things they couldn't fix coped by using acceptance or distractions. Some persons with prHD who had given up used avoidance strategies such as social withdrawal.

Not Broken Yet

Participants who didn't notice changes or who stated changes were not severe did not need to use any coping strategies: "In my head…I'm only carrying the gene; it [has] not come into my lifestyle yet" [P22];

> Every now and then I'll forget [to do something]….I guess it doesn't worry me excessively. I just try to keep track of [it]….And if it happens a lot, I would start to worry about it. But it doesn't seem to be a lot. It's just occasional [P05];

Companions also stated there were no changes yet and thus they didn't need to cope: "The situation isn't bad, so I don't feel the need to resolve anything" [C02];

> The only thing we have to deal with is that he has [the gene]. Thank goodness we don't have to deal with symptoms also right now [C07].

One companion stated she didn't think the interview was relevant to the couples' situation at all because her husband wasn't having any symptoms yet. She said there were more relevant questions related to coping in prHD:

> I think some of those responses would be vastly different if [my husband] was symptomatic and it was affecting our lives….I

> think there are other issues,…like how do I deal with the fact that he doesn't really want to talk about it with other people, but I might want to have support from my friends and might want to talk about it? [C10].

Coping and HD Attributions

Coping strategies did not seem to differ according to whether participants attributed changes to HD or not. Instead, strategies were directed toward specific stressors regardless of the attributions. For example, it didn't matter whether persons with prHD attributed memory problems to HD or to aging; they coped mostly by using memory aids. Persons with prHD who did attribute changes to HD were more likely to use avoidance strategies, such as social withdrawal or not doing tasks that were problematic like driving or doing new things. Companions who attributed changes to HD used more coping strategies than companions who didn't notice changes or attributed changes to things such as aging or temperament. Companions who attributed changes to HD used active coping strategies for things they thought they could fix and acceptance or distraction for those they couldn't.

Comparison of Quantitative and Qualitative Coping Analyses

Side by side comparison of coping strategies endorsed by participants on the Brief COPE as well as in the interviews is presented in Table 8. In the interviews, participants described coping strategies similar to those that appear on the Brief COPE as well as some that do not. The three most commonly used coping strategies that were described in the interviews by persons with prHD were: active coping (N=16; 69.6%), instrumental support (N=12; 52.2%), and use of prescription drugs (N=9; 39.1%). While instrumental support is usually conceptualized as pertaining to tangible support (Langford, Bowsher, Maloney, & Lillis, 1997), in the Brief COPE the items for instrumental support include: "getting help and advice from other people," and "trying to get advice or help from other people about what to do," while emotional support referred to getting emotional "support,

comfort, or understanding" from others (C. S. Carver, 2007). Therefore, in order to provide consistency between the quantitative and qualitative measures, comments were coded as instrumental support if participants stated they sought advice, assistance, or information.

Persons with prHD described all of the coping strategies included on the Brief COPE except denial and self-blame. All persons with prHD stated on the Brief COPE that they used emotional social support. In most cases, persons with prHD told the interviewer that the person from whom they obtained emotional support was their partner. However during the semi-structured interviews, persons with prHD rarely talked about emotional support. Four coping strategies mentioned by persons with prHD that are not on the Brief COPE were: use of prescription drugs (N=9; 39.1%), dyadic coping (N=4; 17.4%), self-monitoring (N=4; 17.4%) and hope (N=4; 17.4%). Persons with prHD used prescription medications to treat depression, anxiety, sleep problems, and distractibility. In all cases, persons using prescription drugs stated they were effective. Dyadic coping refers to coping as a couple, specifically talking about changes together and seeking help as a couple:

> Well, normally when I notice stuff or my husband notices stuff, we talk it out…[H]e'll say, "I notice this and I notice that. What do you think?" [P06];
>
> [I]f either of us gets out of sorts with the other one and feels the need to go see [the psychologist], we go together [P01];
>
> [W]e really help each other in a lot of areas, you know. We assist each other and, you know, pay attention to each other's things [P15].

Some persons with prHD self-monitored for changes in order to control them better: "I truly feel anger….So I have a little bit of concern there….And I will watch it" [P01]; "I'm starting to pay attention to my body and noticing things like being dehydrated and how that affects my lethargy" [P12]. Three persons with prHD described the hope that they will be able to control changes: "I'm hoping that if I really start not being this

normal guy that I've always been…hopefully I'll be able to grasp it before it's too late" [P07];

> But you know that's the thing I'm hopeful about is [that my mother with HD] didn't know the issues, like how much blood sugar affected her….I have to…stay on top of my need for food, so that those things [fighting with her partner] don't happen [P15];

> [T]he other thing that has given me a lot of hope and encouragement is there's also been a lot of things on brain plasticity…. I can have some control over how I train my brain [P12].

One person with prHD hoped changes were not related to HD "because really in my head I don't have it yet" [P23]. The most commonly used coping strategies described by companions in the interviews were active coping (N=8; 34.8%), acceptance (N=6; 26.1%), and helping partners (N=5; 21.7%), discussed above. A companion hoped his partner would "make it" until she was able to qualify for full insurance benefits to cover future nursing home costs [C19]. Hope and helping partners were not strategies on the Brief COPE.

In the qualitative analysis, persons with prHD described using more coping strategies than companions, which is consistent with the results from the Brief COPE. While persons with prHD used self-monitoring to anticipate and control problems, two companions expressed concern that their partners focused on HD too much, which inhibited them from leading normal lives. For example, one companion talked about his wife's "worsening depression" and suicide attempt [C20]. While she attributed her depression to "early onset Huntington's," her husband seemed frustrated by her focus on being "gene positive for Huntington's." He attributed her depression to "an inability to handle the day to day life stresses of raising a family." Another companion was also frustrated with his wife's inability to enjoy normal life due to her focus on HD:

> [T]here appears to me to be no symptoms…but she sees them starting to manifest….[B]ut I think…in some way she uses that as shock value to kind of wake me up to, "Hey, we need to do something and we need to do it now because, you know, next week I'm gonna be laying there in a vegetative state…and our lives will

> gone.”….[T]o her…it just feels like her clock is ticking….And…that to me…is what is robbing her of her joy and her quality of life [C04].

Discussion

The low total mean scores on the Brief COPE for both persons with prHD and companions indicates that participants did not use these coping procedures very often since 1 equals “not at all” and 2 equals “a little bit.” Either they did not need to cope often or they used coping strategies that were not on the Brief COPE. The qualitative findings support the former since the majority of participants stated they had very little to cope with, and participants used only a few coping strategies that were not on the Brief COPE. Lazarus and Folkman’s conceptualization of coping as an imbalance between demands and resources may be an appropriate conceptualization of coping in prHD since these results suggest demands were not currently exceeding resources. However, some participants had noticed more changes and were using more coping strategies, suggesting that it is important to assess coping longitudinally in this population.

The three most frequently used coping strategies in this study are similar to those used by persons with diagnosed HD and their spouses using the COPE scale (Helder, Kaptein, Van Kempen, Weinman, Van Houwelingen et al., 2002a; Helder, Kaptein, Van Kempen, Weinman, Van Houwelingen et al., 2002b). In these studies, participants used acceptance, active coping and planning most frequently. One difference in the current study is that persons with prHD used emotional support as one of the top three coping strategies as reported on the Brief COPE. Participants did not talk about seeking emotional support in the interviews, however. This may be because the interviews focused on concrete changes and how persons coped with them. This also raises the issue that research participants are more likely to endorse things they are probed to endorse.

Most participants noticed subtle changes but attributed them to aging, temperament, and other stressors, which is consistent with previous findings (Downing et al., 2010). This is also consistent with the CSM literature which states people tend to

attribute subtle and ambiguous changes to more benign conditions and wait until symptoms are severe and distinctive before attributing them to illness (H. Leventhal et al., 1998). Mean age of persons with prHD in this study (48.96; SD=11.8) was slightly higher than mean age of all participants in the PREDICT-HD study (43.95; DS=10.33). Thus it was not surprising that some participants attributed subtle changes such as memory problems to age. On the other hand, given that the average age of onset of HD is between 35-55 (Quarrell, 2008), it was also surprising that more participants didn't attribute at least some changes to HD. This appears to violate the symmetry rule of the CSM which states that persons with an illness label will attribute noticed changes to that illness. In the current study, companions and persons with prHD were equally likely to attribute changes to HD. This is in contrast to a prior study that found partners were more likely to notice HD-related symptoms (Kaptein et al., 2007).

The low incidence of HD attributions might indicate participants used denial as a coping strategy. Although denial was one of the least used coping strategies endorsed by participants on the Brief COPE, denial is considered an unconscious defense mechanism (Cramer, 2000). Thus, it is conceptually improbable that participants would endorse it on a questionnaire. Research participants with other illnesses also report low frequency of denial and high frequency of acceptance (de Tychey, Spitz, Briancon, Lighezzolo, Girvan, Rosati, et al., 2005; Llewellyn, McGurk, & Weinman, 2007; Vosvick, Koopman, Gore-Felton, Thoresen, Krumboltz, & Spiegel, 2003).

The most frequently used coping strategy by both persons with prHD and companions in the current study was acceptance. Because persons with prHD in this study had independently obtained HD testing and participated in HD-related research, it is reasonable to assume they have accepted their condition. Nevertheless, the tendency for participants to attribute changes to things other than HD in the interviews suggests they may be using denial as a defense mechanism. While many of the changes noticed by participants were subtle, a growing body of research indicates that many of them may be

related to HD. For example, persons with prHD estimated to be ≤15 years from motor diagnosis performed significantly worse than controls on half of 19 cognitive tests administered; those estimated to be ≤9 years from diagnosis scored worse than controls on almost all of the tests (Stout, Paulsen, Queller, Solomon, Whitlock, Campbell, et al., 2010).

Denial is not necessarily a negative coping strategy; it can be helpful in coping with an illness that is severe and has a poor prognosis (Lazarus, 1999), which certainly pertains to HD. Avoidance can also be considered a form of denial (Seiffer, Clare, & Harvey, 2005). In the current study some participants actively avoided problematic issues instead of trying to fix them. Thus it is difficult to characterize avoidance as a negative or a positive coping strategy. One would have to measure outcomes, such as quality of life, or relationship satisfaction in order to determine the relationships between denial and avoidance and wellbeing.

Another way to characterize denial which may be easier to view as a positive coping strategy is to view it as normalization (Deatrick, Knafl, & Murphy-Moore, 1999). Normalization occurs when people living with chronic illness attempt to construct their lives as normal. When persons first learn about an illness, normal life is disrupted, but over time they can adjust and view their lives similarly to people who don't have the illness (Robinson, 1993). They work, raise children, and socialize. While this can have a positive effect by allowing people with chronic illnesses to experience life as normal, it can have negative consequences if people minimize problems to the extent that they fail to take action when it might be beneficial. A few participants in the current study were making plans for the future by ensuring finances would be sufficient or making career choices that would allow persons with prHD to work longer and/or enjoy their personal time. However, it is possible that if persons with prHD and their companions normalize life too much, they may not adequately plan for the future or fail to notice changes that might have important consequences, such as parenting. For example, persons who grew

up in families with HD-affected parents reported high rates of family dysfunction (Vamos, Hambridge, Edwards, & Conaghan, 2007). It is possible that earlier recognition of changes, especially mood and behavior changes that impact family functioning may alert persons to the need to make changes, such as rearranging roles or altering their methods of communication. Family counseling may be necessary to prevent long-term negative consequences.

Another possible explanation for why persons with prHD in this study did not attribute changes to HD is because of diminished insight that accompanies brain changes in persons with the HD gene expansion (Duff, et al., 2010a). The concept of diminished insight and its relationship to denial in prHD was discussed more extensively in Chapter 3.

The possibility of diminished insight does not explain why companions attributed changes to HD at the same rates, however, which points to other processes at work perhaps in addition to diminished insight. Companions of persons with prHD also have a lot to lose when their partners become severely affected by HD. Companions experience disruption in the work lives and their social lives when partners begin to have severe symptoms (McCabe, Roberts, & Firth, 2008). Thus, they may choose denial or normalization as coping strategies rather than accepting their partners are affected by HD before they may be ready to accept this. Some researchers have proposed there is a type of personality more prone to denial of illness symptoms (Weinstein, Friedland, & Wagner, 1994). Persons with early Alzheimer disease who used denial tended to be focused on work and were very organized, and controlled. Persons with prHD in the current study may have some of these tendencies because they chose to have HD testing, which was perhaps a way to have some control over their disease (Klitzman, 2010). Family members of persons with progressive cognitive illness may also take the affected person's lead in denial of symptoms (Hirschman, Kapo, & Karlawish, 2006). While participants may be using denial or normalization, another possibility to explain the low

incidence of HD attributions is that persons with prHD and their companions do not know what changes to expect prior to HD diagnosis. This possibility was discussed more extensively in Chapter 3.

Persons with prHD in this study used coping strategies more often than companions as measured by both the Brief COPE and the interviews. However, females used coping strategies more frequently than males on the Brief COPE, indicating possible gender bias. Because approximately 74% of the persons with prHD in this study were female, it is impossible to determine whether differences in frequency of use of coping strategies are related to being a person with prHD or gender. In qualitative studies it is not necessary to have equal numbers of males and females because the purpose is to find rich sources of information, regardless of demographic qualities (Chang, Voils, Sandelowski, Hasselblad, & Crandell, 2009). However, the results of the quantitative portion of the current study suggest gender may impact coping, a difference that should be noted even in qualitative analyses.

Persons with prHD did not notice more changes than companions; however, persons experiencing the changes directly logically may need to use more frequent coping strategies. Often the coping strategies described by companions related to reacting to partners' changes, including how they responded to partners' irritability, and helping partners when they noticed the need. In this study, Brief COPE results indicated companions used acceptance, planning, and active coping most frequently. This is similar to previous findings of companions of persons with diagnosed HD using the longer COPE scale (Helder, Kaptein, Van Kempen, Weinman, Van Houwelingen et al., 2002b). In that study, companions used acceptance, positive reinterpretation and growth, active coping, and planning. An important difference is that companions of persons with diagnosed HD used positive reinterpretation and growth, while in the current study companions did not use positive reframing often.

The interview data identified some coping procedures that are not present on the Brief COPE, either because they were specific to prHD (self-monitoring for HD-related changes and hope they will be able to control behavior) or they pertained to coping as a couple (dyadic coping and helping their partners). Several persons with prHD coped by using prescription drugs. All of the participants taking prescription drugs stated they were helpful. This coping strategy was not captured on the Brief COPE. The low frequency of substance use indicated by participants on the Brief COPE (Carver, 2007) indicates they did not consider using prescription drugs to constitute substance use. Persons with prHD who experience depression, anxiety, or sleep disruption may benefit from using prescription drugs to treat these symptoms. Notably, it was not necessary for participants to attribute these symptoms to HD in order to benefit from prescription drugs.

The coping strategies that were used most frequently by participants—acceptance, planning, and emotional support—suggest that participants accepted their situations and used both problem-focused (planning) and emotion-focused (acceptance and emotional support) coping strategies. Although prior studies indicate women used more emotion-focused coping than men (Tamres et al., 2002), in this study both persons with prHD (mostly female) and companions (mostly male) stated they used active coping more than other strategies both on the Brief COPE and in the interviews.

Limitations

This study had several limitations. First of all, the Brief COPE may not be appropriate for assessing coping in prHD. Participants were able to provide feedback on their responses because the scale was administered orally. Several participants found it difficult to respond to the scale because they did not notice changes that significantly interfered with functioning. One participant stated there are other issues related to living with prHD that might be more relevant than how they are coping with changes. However, a few couples did notice changes that interfered with functioning. Therefore, it cannot be

assumed that all persons with prHD do not need to use coping strategies and are coping well. It is also possible that persons who did not respond to invitations to participate in this study were experiencing more severe changes than those who participated, including depression, fatigue, apathy, and social withdrawal, which kept them from participating.

The results of this study suggest the Brief COPE was also not appropriate for assessing dispositional coping in prHD. Three items in the scale, for example, refer to a "situation;" thus, some participants commented they didn't have a "situation" with which to cope at that time. Other coping researchers have argued you cannot assess dispositional coping in a cross-sectional study because it is necessary to measure coping over time and in a variety of contexts and look for consistency in strategies (Lazarus, 2000).

While all participants are living in the context of prHD, many were also coping with other life stressors, including parenting, extended family issues, finances, and moving. Therefore, some participants had these stressors in mind when they completed the Brief COPE and not necessarily HD. Variations in the use of coping strategies may thus be related to types of stressors rather than differences in prHD coping styles. Participants in this study were also at different family development stages. For instance, some were recently married without children, some had young children, some had grown children and grandchildren, and some were retired. The differences in these developmental stages may impact how people cope (Brouwer-DudokdeWit, Savenije, Zoeteweij, Maat-Kievit, & Tibben, 2002). In the current study, the sample size was too small to explore differences between persons at different family development stages.

Another limitation of this study is that there were no outcome measures. Thus, it is impossible to evaluate whether attributions and coping strategies were related to wellbeing. The Brief COPE may be more meaningful when used to explore the modulating influences of coping strategies on outcomes rather than looking only at how often participants used each strategy.

Ideally, the researcher would have liked to recruit equal numbers of females and males due to potential gender differences in coping. Prior studies indicate there may be gender differences in the ways people cope (Galdas, Cheater, & Marshall, 2005; Neff & Karney, 2005). In the PREDICT-HD study, 63.4% of participants with prHD are female, so it would be necessary to use purposeful recruitment in future studies to obtain more equal number of females and males.

Finally, the couples in this sample may not be representative of most couples living with prHD. For example, the longevity of couples' relationships in this sample was notable. Most couples had known each other a long time (median=21 years). This is a strength in the sense that companions would be more likely to recognize changes in their partners. Indeed, persons who had known their partners for the least amount of time had difficulty stating whether partners' behaviors were changes or their personalities or temperaments. On the other hand, the longevity of relationships in this study may not be typical of most couples affected by HD. An older study indicated the divorce rate in couples in which one partner has HD is not higher than in the general population (Tyler et al., 1983). However, marital breakdown most likely occurred within two to three years of disease onset and was associated with more severe behavior changes in the person with HD. The researchers were unable to find more recent data on divorce rates in HD; however, a study of marital relationships in neurological diseases indicated couples with a partner affected by diagnosed HD experienced greater marital dissatisfaction than couples with a partner affected by Parkinson's disease or multiple sclerosis (O'Connor, 2008).

Most of the couples in the current study did not report changes that severely interfered with functioning; thus, relationships that may be challenged by changes in the future were not strained at that point. Couples who did report changes that interfered with functioning reported more relationship problems than those with fewer or less severe changes. There may differences in couples who participate in research together as well.

They may have a greater level of cohesiveness than couples who don't participate in research together. Moderate correlations between religious coping (r=0.51) suggest these couples share similar values that may contribute to greater cohesiveness.

Participants in this study may also differ from other persons with prHD because they have been tested for the HD gene expansion while most people at risk for HD still forgo genetic testing (Tibben, 2007). Persons at risk for HD who predicted they would not cope well with test results may be less likely to undergo testing (Codori et al., 1994). Persons who do not undergo HD genetic testing have been shown to use more avoidance, self-blame and wishful thinking than persons who do (Pakenham, Goodwin, & MacMillan, 2004). Thus, there may be important differences between how people cope depending on whether they undergo HD genetic testing and participate in research or not. It is also possible that persons with prHD who were experiencing more symptoms did not respond to the invitation to participate.

Implications

The results of this study may provide useful information to assist couples in coping with prHD. Participants who used prescription drugs, for example, stated they were helpful, particularly in treating depression. Depression has been associated with reduced health-related quality of life in persons with HD (Ho et al., 2009). Couples who used instrumental support, including therapy and seeking information about HD also stated these coping strategies were effective. While neither of these are cures for HD, they may help people cope with daily life.

Researchers who explored illness representation in inherited cancers concluded participants' illness representations could be used to design cognitive interventions to improve coping (van Oostrom et al., 2007). Similar interventions may be useful in helping persons with prHD and companions cope with changes in prHD. While current research demonstrates researchers can identify cognitive changes in persons with prHD

years before onset (Paulsen, 2010), it is still difficult to state definitively that changes participants noticed in the current study are related to HD. However, some interventions based on the CSM may help persons with prHD and companions cope better. Couples' counseling to assess for problems related to differences in partners' illness representations may improve coping for both partners (Heijmans, De Ridder, & Bensing, 1998; Sterba et al., 2008). Interventions with couples to evaluate and alter illness representations have improved coping and outcomes in other illness processes (Broadbent, Ellis, Thomas, Gamble, & Petrie, 2009a; Keogh et al., 2007).

It was not necessary for participants to attribute changes to HD in order for them to take action or seek help. Instead, participants focused on things they could change and took action to fix them. However, participants who reported changes that interfered with functioning who attributed them to HD talked about acceptance and self-distraction coping strategies in the interviews. Therefore, couples may benefit from knowing early changes may be related to HD so they don't try to fix things that cannot be fixed. Acceptance of changes by both persons with diagnosed HD and their partners has been associated with better mental health in persons with diagnosed HD (Helder, Kaptein, Van Kempen, Weinman, Van Houwelingen et al., 2002a; Kaptein et al., 2007).

Future research with larger samples and more equal numbers of male and female persons with prHD may help sort out which coping strategies are related to gender. Longitudinal data would provide greater accuracy in distinguishing between situational and dispositional coping as well as how couples cope as they encounter more HD-related changes. These data would be useful in developing interventions to enhance coping in persons with prHD and their companions. Future studies should also include outcome measures of wellbeing, such as quality of life or relationship satisfaction. Previous quantitative studies in diagnosed HD using illness representation measures, the Brief COPE and quality of life measures have yielded interesting relationships between coping

and quality of life (Helder, Kaptein, Van Kempen, Weinman, Van Houwelingen et al., 2002a; Helder, Kaptein, Van Kempen, Weinman, Van Houwelingen et al., 2002b).

In the prHD population, it would be helpful to understand whether attributing changes to HD influences coping strategies and how it affects wellbeing. Regression analyses using the Brief COPE as a situational measure of coping with prHD may reveal differences in coping strategies between people who attribute changes to HD and those who do not as well as the mediating and moderating relationships between coping strategies and measures of wellbeing. Using an outcome measure would also allow for analysis of dyadic coping by exploring how each partners' coping strategies affects outcomes in the other (Berg & Upchurch, 2007).

There are no data currently that explore whether persons with prHD and their companions want to learn that changes they notice may be related to HD. Since the persons with prHD in the current study participate annually in longitudinal research of prHD, it is theoretically possible to offer them their personal study results. This would enable them to see their patterns of change over time as well as the areas of functioning that are in decline. This information may be helpful for making future plans and adjusting roles. For example, several couples in the current study had adjusted their roles by allowing their spouse to take over more housework, cooking, and shopping. On the other hand, the perceived risks may include increased depression and hopelessness and premature role adjustment, such as work cessation. Future studies are necessary to explore perceived risks and benefits of receiving this information from the perspective of persons with prHD and their companions.

Finally, there are issues couples living with prHD may be coping with that might be more relevant than focusing on functional changes, especially before changes impact daily life. For example, a companion in this study mentioned the stress she feels related to not being able to get support from other people because her husband doesn't want anyone to know his HD gene status. This is an issue that might be helpful to study in

order to better address this concern. It is important to ask persons with prHD and their companions about other issues they would like help with that have not yet been explored.

Table 7. Most and Least Frequently Used Coping Strategies on the Brief COPE

		Persons with prHD		Companions	
	Coping Strategy	**Mean**	**SD**	**Mean**	**SD**
Most frequently used coping strategies	Acceptance	3.29	0.48	3.18	0.90
	Emotional Support	3.23	0.67	2.56	1.11
	Planning	2.92	0.74	2.48	0.78
Least frequently used coping strategies	Denial	1.20	0.36	1.29	0.45
	Substance Use	1.39	0.65	1.15	0.53
	Behavioral Disengagement	1.27	0.39	1.15	0.41

Table 8. Comparison of Quantitative and Qualitative Coping Strategies

Coping Strategy	Brief COPE		Interview	
	PrHD Number (%) who used strategy at least "a little bit"	**Companion** Number (%) who used strategy at least "a little bit"	**PrHD** Number (%) who mentioned strategy	**Companion** Number (%) who mentioned strategy
Emotional Support	23 (100%)	17 (74%)	2 (8.7%)	1 (4.3%)
Acceptance	22 (95.7%)	22 (95.7%)	4 (17.4%)	6 (26.1%)
Active Coping	22 (95.7%)	20 (87%)	16 (69.6%)	8 (34.8%)
Instrumental Support	22 (95.7%)	17 (74%)	12 (52.2%)	2 (8.7%)
Venting	20 (87%)	17 (74%)	3 (13%)	0
Planning	19 (82.6%)	20 (87%)	6 (26.1%)	2 (8.7%)
Positive Reframing	19 (82.6%)	19 (82.6%)	1 (4.3%)	1 (4.3%)
Self-Distraction	18 (78.3%)	15 (65.2%)	4 (17.4%)	4 (17.4%)
Religion	14 (60.9%)	14 (60.9%)	1 (4.3%)	0
Self-Blame	10 (43.5%)	10 (43.5%)	0	0
Humor	9 (39.1%)	9 (39.1%)	2 (8.7%)	0
Behavioral Disengagement	5 (21.7%)	7 (30.4%)	5 (21.7%)	4 (17.4%)
Denial	3 (13%)	3 (13%)	0	0
Substance Use	3 (13%)	3 (13%)	1(4.3%)	0
Prescription medications	na	na	9 (39.1%)	0
Dyadic Coping Strategies	na	na	4 (17.4%)	0
Helping partner	na	na	Na	6 (26.1%)
Hope	na	na	4 (17.4%)	1 (4.3%)
Self-Monitoring	na	na	4 (17.4%)	na

CHAPTER 5

DISCUSSION AND CONCLUSIONS

Summary of Findings

The purpose of this study was to explore illness representations and coping procedures of persons with prHD and their companions. Qualitative methods were used to explore illness representations using the CSM as a framework, while a mixed methods approach was used to explore coping. Twenty-three couples participated. The following specific aims were addressed in three papers:

1. Describe the attributions that persons with prHD and their companions made for functional changes in prHD and the active processes used to make attributions;
2. Explore illness representations in persons with prHD and companions using CSM and evaluate the appropriateness of the CSM in anticipated illness using prHD as a model;
3. Describe the coping strategies used by persons with prHD and their companions to manage changes.

A summary of each paper and their findings are presented below, followed by a discussion of overall study findings, limitations, and clinical and research applications.

Paper 1

The first paper presented findings from a study that explored illness representations in eight persons with prHD and seven of their companions. Data came from semi-structured interviews conducted to explore changes in work function in prHD. The CSM was used as a framework to conduct analyses. Results of this study indicated persons with prHD and companions used the CSM processes: Participants noticed changes in work function and some noticed changes in other areas of functioning; most

also made attributions for changes although they were not asked to do so; they demonstrated the use of active processes in deciding to what they attributed changes.

Specifically, participants noticed changes in memory, mood, and behavior. Behavior changes included irritability and stubbornness. Work function changes included issues with coworkers, receiving negative reviews, and feeling overworked. Other changes included being late and obsessing about things. Participants attributed changes to aging, temperament, work issues, or other health conditions. Active processes included comparing the person with prHD to others with and without HD, and symptom monitoring. Many participants expressed uncertainty regarding how to make attributions for noticed changes, and some wondered whether some changes were related to HD. The symmetry rule of the CSM was not supported because only one participant attributed changes to HD. Results of this study were considered preliminary, however, because they came from a secondary analysis of qualitative data that were not collected to explore illness representations.

Paper 2

The purpose of the second paper was to build on the findings of the first paper by purposefully exploring illness representations in prHD using the CSM in a larger sample. A second aim was to evaluate the appropriateness of the CSM in anticipated illness using prHD as a model. The focus was expanded beyond making attributions and using active processes to include the other elements of the CSM, including using and evaluating coping strategies to address changes. Twenty-three couples participated in semi-structured interviews. Two of these couples had also participated in the study presented in Paper 1. Thus they were interviewed again approximately one year later.

Data were analyzed using descriptive analysis. Results supported preliminary findings: Participants noticed functional changes, made attributions, and demonstrated active processes in making attributions. In addition, they used coping strategies when

changes impacted functioning, and evaluated their effectiveness. However, some processes did not go beyond noticing changes. Several participants noticed changes but said they didn't know what they were related to or they couldn't decide whether they were related to HD or to other things. Again common attributions were aging, temperament, and other health conditions. In addition, they attributed changes to other life stressors. Participants did not need to make attributions in order to use coping strategies and evaluate them, suggesting the link between illness representations and coping in prHD is tenuous.

In this study, 14 participants attributed changes to HD: Seven persons with prHD and seven companions. Four couples were congruent in attributing changes to HD while three persons with prHD and three companions attributed changes to HD when their partners did not. Incongruent couples mentioned more relationship issues. As in Paper 1, the symmetry rule was only partly supported because while participants made attributions for changes, less than a third attributed changes to HD.

Implications for this study include the possible benefit of assessing the illness representations of persons with prHD and their companions. While it was not necessary for participants to make attributions in order to cope effectively, incongruent illness representations could contribute to relationship issues. Further research that includes outcomes could provide greater insight into the link between illness representations and coping in prHD.

Paper 3

Paper 3 presents findings from a mixed-methods study conducted to explore coping in persons with prHD and their companions. While little is known regarding whether persons with prHD and companions notice changes in prHD and attribute them to HD, even less is known regarding how they cope with changes. This study was conducted to address that gap.

The quantitative measure used in this study was The Brief COPE (Carver, 2007). Results indicated persons with prHD used acceptance, emotional support, and planning more frequently than other coping strategies. Companions used acceptance, planning, and active coping. Both partners used denial, substance use, and behavioral disengagement least frequently. Persons with prHD used coping strategies more often than companions. However, overall coping scores indicated participants did not use coping strategies frequently. This was supported by verbal feedback during administration of the scale that most participants felt as though they didn't need to cope with HD yet. This suggests that current demands were not exceeding current resources (Lazarus, 1999) for most participants.

Three themes were identified in the qualitative analysis: trying to fix it, can't fix it, and not broken yet. Participants used active coping strategies to cope with changes they appraised as fixable. For example, the majority of persons with prHD used visual reminders to cope with memory changes. Persons with prHD as well as companions used strategies to prevent or mitigate irritability in persons with prHD. Some changes were appraised as not fixable, including social withdrawal, apathy, and clumsiness. Participants either coped by accepting things they couldn't fix or avoiding them. Finally, participants did not use coping strategies when they didn't notice any changes or when changes were not severe enough to interfere with functioning.

The results from the qualitative analysis suggest persons with prHD use strategies to cope with changes when changes interfere with functioning. When coping strategies are not effective, they either accepted or gave up. In many cases it did not seem to matter what participants attributed changes to in order for them to cope effectively. However, participants who attributed changes to HD were more likely to use acceptance or distraction to cope. Therefore, the tenet of the CSM that links illness attributions to coping was only partially supported.

The results of this study were limited by possible gender effects in coping since 17 of the 23 persons with prHD were female. The absence of outcome measures also limited analyses. The addition of outcomes measures would strengthen analyses of the relationships between attributions, coping, and quality of life in prHD. It would also allow for analysis of dyadic coping by examining how each partner's illness representations and ways of coping affect the other's quality of life of life.

Nevertheless, this paper represents the first exploration of coping with daily functional changes in prHD. Results may be useful in helping persons with prHD and companions use strategies to facilitate effective coping. There was limited support that attributing changes to HD facilitated effective coping by using prescription medications to treat some changes, and fostering acceptance and self-distraction when changes couldn't be fixed.

Discussion and Reflection

In this section I will discuss and reflect on overall findings, limitations, clinical implications, and suggestions for future research.

Combined Findings

The combined findings of this dissertation study provide a first glimpse into how persons with prHD and companions make sense of and cope with changes between the time of genetic testing and diagnosis of HD. Several studies have explored the impact of HD genetic testing on both persons at risk for HD and their spouses (Tibben, Timman, Bannink, & Duivenvoorden, 1997; Timman, Roos, Maat-Kievit, & Tibben, 2004), using measures of coping and wellbeing. However, little was known prior to the current study regarding whether persons with prHD and their companions noticed changes in daily functioning or how they made sense of and responded to these changes.

These study results support the use of the CSM to explore illness representations in prHD. The elements of the CSM were apparent in the data—participants noticed

changes, made attributions for changes, demonstrated use of active processes to make attributions, and used and evaluated coping strategies. However, the CSM did have some limitations in this sample. For example, in the first two studies participants made attributions, but they did not explicitly talk about the five attributes of illness representation—identity, cause, timeline, consequences, and cure/controllability. In Paper 1, participants volunteered attributions, while in Paper 2 they were asked to make attributions. Attributions could be related to either identity or cause, but due to the overlap of these concepts I chose the term attributions instead. Timeline, consequences, and cure/controllability may have been implicit in their attributions, coping strategies, and evaluations but they did not talk about them directly.

There was also limited support for the symmetry rule: While a few participants looked for changes that might be related to HD, the majority of participants did not attribute changes to HD. The lack of reference to the five attributes and low frequency of HD attributions could be accounted for either by the absence of changes that definitively suggested HD or because participants did not have enough information about changes in prHD to form illness representations.

Not all participants used every element of the CSM. For example, several did not make attributions for noticed changes and this did not prevent them from using coping strategies. This suggests that contrary to the tenets of the CSM, there was not a definite link between attributions and coping strategies. It is possible that beliefs about changes might be coping mechanisms themselves. This is what the authors of the CSM refer to as emotional processing that runs parallel to cognitive processing in the CSM (H. Leventhal et al., 1998). However, it is very difficult to measure emotional processes. For example, "positive reframing," "venting," and "denial" are coping strategies on the Brief COPE (Carver, 2007) that might represent emotional processing. However, participants rarely endorsed using these strategies. This does not mean they did not use these strategies,

though, because they may be ego defense mechanisms that are subconscious and therefore not measurable (Cramer, 2000).

Despite the limitations of the CSM, there were indications from the interviews that some people formed strong representations of prHD, as illustrated by the disappointment of one participant when a repeat test revealed he had fewer CAG repeats than on the first test. This participant's response suggested he had formed a strong illness representation based on his CAG repeat number.

Findings are relevant to components of other illness perception models. For example, the Family System Illness model (Rolland, 1987) uses a developmental perspective which would be appropriate in prHD. In this model there are three time phases: the crisis phase following diagnosis, the chronic phase, and the terminal phase. The task of persons in the chronic phase is to "maintain the semblance of normal life" (p. 4). This phenomenon may explain why most participants in the current study did not attribute changes to HD and normalized their lives. In terms of Erikson's (1994) developmental stages, persons in midlife are focused on generativity, including raising children, accomplishing career goals, and caring for aging parents. Participants in this study were all involved in these activities.

Rolland and Williams (2005) adapted the Family System Illness model for use in genetic illnesses, creating the Family System Genetic Illness model. In this model there are two crisis phases—the first is when persons at risk for genetic illness contemplate testing, and the second during and immediately after testing. Crisis phases are followed by long-term adaptation in which persons balance "open communication" and "proactive planning" with the "need to live a 'normal' life" (p. 16). This model supports findings in the current study that indicated persons with prHD and their companions attempted to normalize their lives. The Family Systems Illness Model may also be more appropriate in anticipated illness than the CSM. Rolland (1990) describes how the model can be used to explore anticipated illness, stating that illness beliefs change over time. This emphasizes

the limitations of cross-sectional studies to characterize illness representations in particular illness because participants may be at very different phases of anticipatory loss.

Another useful model for exploring dyadic coping in illness is the developmental-contextual model proposed by Berg and Upchurch (2007). This framework facilitates dyadic coping analysis, which was limited in the present study by lack of outcome variables. The developmental-contextual model takes into account sociocultural and historical contexts as well as age and gender of couples and marital quality and illness condition focusing on lifespan dyadic appraisal and coping. Illness representation is one component of dyadic appraisal in this model. Thus, it is an ambitious model that requires multiple measures at multiple data collection periods. Nevertheless, the focus on the interaction of partners' appraisal, coping, and adjustment provides a systematic way to assess dyadic coping rather than merely parallel coping.

Findings from the present study support the need to use outcome measures to explore congruence in illness representations and coping between partners and to help identify participants who were distressed. The addition of outcomes measures, such as quality of life, mental health, or life meaning may provide useful information regarding the relationship between illness representations, coping strategies, and well being.

One planned analysis—exploring the relationship between having children and coping strategies—was not conducted for several reasons. The most important reason was that only two persons with prHD did not have children. Beyond this, the issue of children was more complicated than just whether or not people had children. For example, several couples said they had children before they know about HD in their family. A few companions were stepparents, and one couple used selective embryo transplant to ensure their children would not have HD. In addition, some couples were experiencing difficulties with their children, which could confound coping related to having children.

Another interesting ancillary finding was that seven persons with prHD said they didn't know their parents had HD until they themselves were adults. Since not all

participants were asked this question, it is unknown whether more participants grew up without knowledge of their HD risk. Nevertheless, some persons who did not know their parents had HD still recalled tumultuous childhoods due to parents' erratic behaviors. Because illness representations are influenced by past experiences with an illness, the varying experiences of these participants with HD points out the possibility that they could have vastly different illness representations even if they experienced similar changes.

In addition to other models that might be useful in exploring how couples cope during the HD prodrome, results from studies of couples' coping with other progressive neurological illnesses may also be informative, such as ALS and multiple sclerosis (MS). In addition, how people cope with illnesses that involve progressive cognitive decline, such as Alzheimer disease, may also be informative. It is important to use caution when applying these results to prHD for several reasons. First, although ALS and MS have genetic components, they are usually not diagnosed until after persons are experiencing symptoms; the participants in this study, however, were tested before they had distinctive HD symptoms. HD also is an autosomal dominant disease, meaning that most persons with prHD have grown up in a family affected by HD and thus have likely formed illness representations of HD. Only 15-20% of cases of ALS are thought to be caused by an autosomal dominant genetic mutation and penetrance is sometimes incomplete (OMIM, 2010a). It is similar to HD because it has adult onset and leads to premature death; however, it is not usually accompanied by the severe progressive cognitive and behavioral changes associated with HD.

Multiple sclerosis also has adult onset and is accompanied by cognitive and psychiatric changes which make it similar to HD. The genetic component of MS is even less clear than in ALS. Although trends in families have been recorded, no genes have been definitively linked with causing MS, although some have been associated with susceptibility to MS (OMIM, 2010c). Persons with MS also experience a different time

course than persons with ALS or prHD with occasional periods of remission and exacerbation. However, studies of persons with ALS and MS and their companions may help to identify important issues and study methods to explore coping in persons with prHD and their companions. Alzheimer disease is like HD in that it involved progressive cognitive decline; however, it is not associated with the physical symptoms which can contribute to perceived stigma in HD. It might be pertinent to explore how perception of stigma relates to coping strategies in HD as well.

The results of this dissertation study indicate the CSM is a useful model to explore illness representation in prHD, although other models and study designs may also be helpful. Research on couples coping with other illnesses may also provide valuable guidance in designing future studies of coping in prHD. Results also indicate that persons with prHD and companions may benefit from interventions that assess illness representations and help persons choose effective coping strategies based on these beliefs.

Overall Limitations

In addition to the limitations of the CSM and the Brief COPE discussed above, the findings in this study are limited by the sample size and sample characteristics. Furthermore, the study design did not allow for testing the relationships between illness representations, coping, and quality of life.

The recruitment goal was 30 couples. I attempted to contact all 103 participants in the pool of persons with prHD identified by the PREDICT-HD 2.0 coordinator. Current telephone numbers were missing for 40 people on the list. Invitations were sent by mail if phone numbers were missing or persons did not answer their telephones after three attempts. Of the 60 mailed invitations to participate, 12 were returned to sender as not deliverable. Thus, 39 persons with prHD were contacted by phone, 28 were eligible to participate, and 16 persons with prHD and their companions completed the study. Several

persons did not meet eligibility criteria, either because they didn't have a spouse or significant other or they had been diagnosed.

There are several reasons why sample size was less than anticipated. In subsequent interviews with persons with prHD for a different study (Downing, 2010, unpublished data), some participants said they didn't like answering the telephone for a variety of reasons—apathy, reluctance to engage in a conversation with strangers or in which they can not anticipate what the caller will ask, and speech difficulties. Thus, they screen calls with caller ID and don't pick up if they don't recognize the number. Some said they picked up because they recognized the area code as the "PREDICT" area code and they were willing to talk to PREDICT researchers. It is thus plausible that persons with prHD who were experiencing more severe changes such as apathy, depression, social withdrawal, or cognitive changes did not want to answer their telephones. However, the low response rate to mailed invitations suggests otherwise since persons had the opportunity to read the materials at their convenience. Reading consent documents and returning signed copies may be overwhelming for persons if they are experiencing cognitive changes or fatigue. Finally, persons with current telephone numbers who answer unknown telephone calls may be more likely to agree to participate in research.

I reached qualitative data saturation after 15 interviews. However, I attempted to reach a sample size of 30 in order to meet Morse's (2009) recommendation of sample size of 30-50 using semi-structured interviews. Another recruitment approach would be to post electronic study invitations on the PREDICT-HD website where many members of HD families know where to find them.

Participants in this study may not be representative of most persons with prHD and their companions. In addition to self-selecting for this study, all potential participants have been tested for the HD gene expansion and have previously participated in HD-related research. The uptake of HD predictive testing is estimated to be less than 25%

(Tibben, 2007). Even fewer people than that participate in HD-related research. Thus, the participants in this study are a unique group of individuals. The couples in this study might have stronger relationships than other couples affected by HD because participating in research together may be a sign of greater cohesiveness. This is supported by the high median number of years partners had known one another (21 years). On the other hand, it is still possible that the coping strategies that were effective for participants might also be beneficial for others who are coping with prHD changes.

Finally, the results of this study cannot tell us anything about the impact of illness representations and coping on quality of life in persons with prHD and their companions. Future studies that include outcome measures are necessary to provide these data. This would also allow for the exploration of the role of coping strategies as mediator and/or moderators of quality of life. Another outcome measure that might be useful to explore is life meaning. In a study involving patients with Amyotrophic Lateral Sclerosis (ALS), persons with ALS shifted life meaning toward relationships and away from health as their disease progressed (Fegg, M. J. et al., 2010). Interventions aimed at strengthening relationships in persons with prHD might be useful for helping them find positive meaning in life.

Clinical Implications

Altering Illness Representations

Several studies have indicated that illness representations can be altered through counseling to facilitate more effective coping and improve quality of life. The findings from this study indicated that for the most part participants were not experiencing significant impairment in functioning. However, for the few who were, effective coping strategies included using prescription drugs, accepting changes that couldn't be fixed, and using distractions when changes couldn't be fixed. These strategies may be useful to other persons with prHD and their companions. The cognitive changes that accompany

HD can lead to behaviors that strain relationships (Williams, Hamilton, Nehl, McGonigal-Kenney, Schutte, Sparbel, et al. (2007); therefore, companions especially might benefit from knowing that changes before diagnosis may be related to HD. They may benefit from support that helps them accept changes they cannot fix and to find ways to distract themselves when they need a break from care giving or thinking about HD.

It might be helpful for healthcare providers to assess illness representations in both persons with prHD and their companions. Even the act of discussing illness representations have been shown to improve patient-physician communication and lead to more effective treatments (de Ridder et al., 2007). Many participants in this study were seeking information—they were reading the HD newsletter, looking up things about HD, asking their physicians about changes. Interestingly, they also sought information from researchers during their participation visits and looked for clues in the study surveys regarding what changes to expect. This suggests that some persons with prHD and their companions want information regarding changes in prHD. In patients with ALS, early diagnosis of the disease provided validation of symptoms they had noticed (Gelinas, 2000). However, not all participants sought information; for many participants life was relatively normal. Changes were due to things "normal" people have such as aging, temperament, or other life stressors. One woman even stated she was "just like every other person that walks the face of the earth."

Disclosure of Research Results

The finding that many participants were trying to find information about prHD changes during research participation raises ethical issues related to the risks and benefits of disclosure of individual research results. On the one hand, the growing divide between what researchers now know about changes in prHD (Paulsen, 2010) and what persons with prHD and companions know, based on the current study, suggest that not informing persons that changes may be related to prHD represents paternalism (Fernandez, 2008).

On the other hand, paternalism may also be demonstrated by researchers' beliefs that withholding information is harmful (Meyer, 2008).

There may be a difference between wanting information about general changes in prHD and wanting to receive individual research results. It is unclear from this study how persons with prHD and their companions would respond if they were told changes might be related to HD. More information is needed regarding whether persons who participate in prHD research want to receive individual research results and how they perceive risks and benefits. If research participants indicate a desire to receive individual results, guidelines for return of results can be modeled on those used in predictive HD testing (Nance, R. Myers, Wexler, & Zanko, 2003). For example, participants should be accompanied by a support person, and counseling should be mandatory and include sufficient time for participants to ask questions and process the information.

Implications for Future Research

The results of this study suggest several avenues for future research. This study provides a first glimpse into how persons with prHD and companions make sense of and cope with changes in prHD. Future research could expand on these findings by including outcome measures to explore the relationships between illness representations, coping, and quality of life. For example, multiple regression models could be used to test for mediation and moderation effects of coping strategies. It would also be helpful to explore whether attributing changes to HD or not impacted coping strategies and quality of life. Thus, whether or not participants attributed changes to HD could be entered into a multiple regression model as a dichotomous variable.

A limitation of using the Brief COPE in multiple regression analyses is that it consists of 14 scales. Therefore, a multiple regression model would require a sample size of at least 135 to accommodate 14 predictor variables with power=0.8, alpha=0.05 and estimated effect size=0.15 (Soper, 2010). Thus, it is understandable why some

researchers have succumbed to the temptation to combine the scales into fewer categories by using factor analysis (Myaskovsky et al., 2005) or sorting them into emotion-focused and action-focused (Cooper et al., 2006) or adaptive and maladaptive (Meyer, 2001) coping strategies. Some coping measures used in quantitative analyses include fewer scales (Hagger & Orbell, 2003), including the Ways of Coping Questionnaire (Folkman & Lazarus, 1988), which contains eight scales. Although fewer scales require fewer subjects, the tradeoff is that they yield less information because they only measure a limited number of coping strategies.

It may only be possible to use the Brief COPE as a measure of dispositional coping using longitudinal studies that explore coping in a variety of contexts and evaluate whether coping strategies remain consistent within individuals (Lazarus, 2000). However, participants in the current study didn't respond well to using the Brief COPE as a dispositional measure since some of the items referred specifically to a "situation." Furthermore, participants in the current study were experiencing other life stressors and responded to the Brief COPE with those stressors in mind. In the future, it might be preferable to use the Brief COPE as a situational measure, asking participants how they cope with the stressor of prHD. This would eliminate the potential of other stressors confounding results and eliminate semantic confusion related to the use of the word "situation." If participants state they are not using any coping strategies related to prHD, this would also be informative.

Qualitative methods may be more appropriate for exploring coping than quantitative methods because coping is part of an active process and is transactional in nature (Coyne & Gottlieb, 1996). In the current study, qualitative methods revealed additional coping strategies that were not included on the Brief COPE. Therefore, mixed methods studies may be the best way to explore coping.

Participants in this study varied in the number and degree of changes they noticed, as well in how they coped with changes, indicating they may have been in different

stages of disease progression. Thus, longitudinal research would provide more precise information regarding how persons evaluate and cope with changes over time as they move closer to diagnosis. I was able to get a glimpse of these possibilities in this study because I had interviewed two couples approximately a year earlier as part of another study. The responses for one couple were similar to their first interview—they still had not noticed any changes. The other couple, however, had changed from attributing changes to aging, personality, and injuries to attributing several changes to HD. What had happened in the year between interviews was that the partner with prHD had started taking a medication as part of a drug trial to treat or delay HD-related symptoms. Both he and his wife believed he received the actual drug and not the placebo because several areas of functioning had improved—he was less irritable and tired, he was able to focus better at home and at work, and he was no longer actively avoiding social situations. This reappraisal is the kind of information that can be best obtained using longitudinal methods.

Longitudinal studies would also help to sort out what changes might be related to HD versus those that might be related to other things. These data are critical in identifying markers of HD progression that can be used to evaluate the effectiveness of future treatments in clinical trials (Paulsen, Wang et al., 2010). Furthermore, some participants may simply prefer to know what changes might be related to HD in order to relieve uncertainty.

Finally, it was disturbing to hear the comments of one companion who stated the questions I asked were not relevant to her situation. While this person did not notice any symptoms in the spouse and thus had nothing to cope with related to prHD at that point, this participant was nevertheless coping with the stress or having to keep the spouse's gene status secret. This companion wanted to seek support from friends but could not. This example illustrates there may be other issues to explore that are more relevant to persons living with prHD.

Conclusions

This study provided a first glimpse into how persons with prHD and companions make sense of and cope with changes in prHD. Participants did notice changes, although they were often subtle and not distinctive enough to attribute them to HD. However, lack of information regarding what changes to expect prior to onset of distinctive HD motor signs might partially explain why participants did not recognize changes might be related to HD. While researchers are aware that changes are present several years before motor onset, little is known regarding whether persons with the HD gene expansion or their companions expect changes. While I have used the term prHD in this study, the term "prodromal HD" is not common usage outside the research world. It is not clear how persons affected by HD would respond to such a term.

Nor is it clear from this study whether making persons with prHD and companions aware that changes they experience may be related to HD would be helpful or harmful. More research is needed to explore whether persons with prHD and their companions want this information, including whether they would like to receive individual research results. Future studies that include quality of life outcome measures and studies that explore what persons with prHD and companions want to know could help illuminate these issues.

APPENDIX A

SEMI-STRUCTURED INTERVIEW GUIDE—PERSON WITH PRHD

Thank you for agreeing to participate in this study. I'm going to ask you questions about how things have been going for you—at work, at home, in your social life, your physical activities, and how your mood has been in the past month. I'll ask you if anything is a change and what you attribute any changes to and how you made those decisions. Do you have any questions before we get started?

First, I'd like to ask you a couple of questions about yourself and your relationship with _____:

What is your age? _____

Do you have any children? _____

After a change is mentioned, proceed to questions 2 and 3 then return to list of domains and proceed in that order so only one topic of change is discussed at a time.

1. First of all, tell me how things have been going for you lately—how have you been feeling and have you noticed any changes in your physical functioning, behavior, mood, or thoughts? *Is this a change?*

Allow participant time to respond. If participant has difficulty responding, continue with interview until all domains have been addressed:

How are things going at home? *Is this a change?*

Probes: How are you getting along with family members? How are things going with your home projects or hobbies? How about childrearing or housework?

How are things going in your social life? *Is this a change?*

Probes: How are things with your friends, extended family?

How has your mood been lately? *Is this a change?*

How have things been going in terms of remembering things like appointments, driving directions, shopping lists? *Is this a change?*

How have things been going when you need to make plans for something in the future, like taking a trip or planning an event? *Is this a change?*

How have things been when you need to learn how to do something new, like putting together something you've bought or doing a new task at work? *Is this a change?*

How are things going in terms of physical activities—exercise, working around the house, physical parts of your job, driving? *Is this a change?*

How are things going at work? *Is this a change?*

Probes: How about getting your work done? Getting to work or appointments on time? Interactions with coworkers/supervisors/clients/customers? Enjoying your job as much as you used to?

2. So, you've noticed that _____ is a change for you. What do you think is the reason for this change? *Probe: Why do you think this change has happened?*

3. How did you decide that this change is related to _____?

Continue until participant has nothing more to add.

4. What have you been doing to cope with this change? *Probe: What have you been doing to manage this change?*

5. What can you tell me about how effective this has been in helping you cope with/manage this change? *Probe: Has it been working for you? Why do you think it has/hasn't been working for you? What do you think may be a better way to manage this change? Why?*

We are now finished with the interview. Before you go I would like to ask you what concerns you may have regarding anything we discussed today.

APPENDIX B

SEMI-STRUCTURED INTERVIEW GUIDE—PERSON WITH PRHD

Thank you for agreeing to participate in this study. I'm going to ask you questions about how things have been going for your _____ [state relationship] —at work, at home, in his/her social life, his/her physical activities, and how his/her mood has been in the past month. I'll ask you if anything is a change and what you attribute any changes to and how you made those decisions. Do you have any questions before we get started?

First, I'd like to ask you a couple of questions about yourself and your relationship with _____ :

What is your age? _____

How long have you known _____? _____

After a change is mentioned, proceed to questions 2 and 3 then return to list of domains and proceed in that order so only one topic of change is discussed at a time.

1. First of all, tell me how things have been going for your _____ lately—how has he/she been feeling and have you noticed any changes in his/her physical functioning, behavior, mood, or thoughts? *Is this a change?*

Allow participant time to respond. If participant has difficulty responding, continue with interview until all domains have been addressed:

How are things going at home for your_____? *Is this a change?*

Probes: How are you getting along with family members? How are things going with your home projects or hobbies? How about childrearing or housework?

How are things going in his/her social life? *Is this a change?*

Probes: How are things with his/her friends, extended family?

How has his/her mood been lately? *Is this a change?*

How have things been going in terms of him/her remembering things like appointments, driving directions, shopping lists? *Is this a change?*

How have things been going when he/she needs to make plans for something in the future, like taking a trip or planning an event? *Is this a change?*

How have things been when he/she needs to learn how to do something new, like putting together something he/she has bought or doing a new task at work? *Is this a change?*

How are things going in terms of his/her physical activities—exercise, working around the house, physical parts of his/her job, driving? *Is this a change?*

How are things going at work for him/her? *Is this a change?*
Probes: How about getting his/her work done? Getting to work or appointments on time? Interactions with coworkers/supervisors/clients/customers? Enjoying his/her job as much as he/she used to?

2. So, you've noticed that _____ is a change for your _____. What do you think is the reason for this change? *Probe: Why do you think this change has happened?*

3. How did you decide that this change is related to _____?

Continue until participant has nothing more to add.

4. What have <u>you</u> been doing to cope with this change? *Probe: What have you been doing to manage this change?*

5. What can you tell me about how effective this has been in helping you cope with/manage this change? *Probe: Has it been working for you? Why do you think it has/hasn't been working for you? What do you think may be a better way to manage this change? Why?*

We are now finished with the interview. Before you go I would like to ask you what concerns you may have regarding anything we discussed today.

APPENDIX C

BRIEF COPE

These items deal with ways you've been coping with the stress in your life in the past month. There are many ways to try to deal with problems. These items ask what you *usually* do to cope with stress. Obviously, different people deal with things in different ways, but I'm interested in how you've tried to deal with stress. Each item says something about a particular way of coping. I want to know to what extent you've been doing what the item says—how much or how frequently. Don't answer on the basis of whether it seems to be working or not—just whether or not you're doing it. Try to rate each item separately in your mind from the others. Make your answers as true FOR YOU as you can. Use these response choices:

1 = I haven't been doing this at all
2 = I've been doing this a little bit
3 = I've been doing this a medium amount
4 = I've been doing this a lot

1. I've been turning to work or other activities to take my mind off things.				
2. I've been concentrating my efforts on doing something about the situation I'm in.				
3. I've been saying to myself "this isn't real."				
4. I've been using alcohol or other drugs to make myself feel better.				
5. I've been getting emotional support from others.				
6. I've been giving up trying to deal with it.				
7. I've been taking action to try to make the situation better.				
8. I've been refusing to believe that it has happened.				
9. I've been saying things to let my unpleasant feelings escape.				
10. I've been getting help and advice from other people.				
11. I've been using alcohol or other drugs to help me get through it.				
12. I've been trying to see it in a different light, to make it seem more positive.				
13. I've been criticizing myself.				
14. I've been trying to come up with a strategy about what to				

do.				
15. I've been getting comfort and understanding from someone.				
16. I've been giving up the attempt to cope.				
17. I've been looking for something good in what is happening.				
18. I've been making jokes about it.				
19. I've been doing something to think about it less, such as going to movies, watching TV, reading, daydreaming, sleeping, or shopping.				
20. I've been accepting the reality of the fact that it has happened.				
21. I've been expressing my negative feelings.				
22. I've been trying to find comfort in my religion or spiritual beliefs.				
23. I've been trying to get advice or help from other people about what to do.				
24. I've been learning to live with it.				
25. I've been thinking hard about what steps to take.				
26. I've been blaming myself for things that happened.				
27. I've been praying or meditating.				
28. I've been making fun of the situation.				

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