Investigating Factors Associated With Offspring's Coping with Huntington's Disease-related stress.

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Abstract: *Objective:* In this study, I investigated (1) how age is related to reported levels of coping with Huntington's Disease-related stress in offspring; (2) how measures of disease progression in parents are directly related to use of coping in offspring; (3) how use of coping in parents is directly related to coping in offspring; and (4) if there is an indirect association of disease progression in parents with the ways that offspring cope with HD-related stress through perceived levels of offspring HD-related stress. *Method:* Adolescent (ages 10 –19) and young adult (ages 20 –29) offspring and their parents with HD (n=40) were recruited from the Vanderbilt University Medical Center Huntington Disease Society of America Level 1 Center of Excellence. Offspring participants in the study completed self-report measures of coping and neuropsychiatric symptoms, and parents completed measures of coping and had their genetic status measured. *Results:* Age was found to have a significant negative correlation with disengagement coping. CAP scores were found to be significantly negatively associated with secondary control coping. Number of CAG repeats was also found to be significantly negatively associated with secondary control coping and was found to be significantly positively associated with disengagement coping. Parents' primary control coping was significantly positively associated with offspring's primary control coping. Parents' disengagement coping was found to be significantly negatively associated with offspring's primary control coping. Perceived levels of offspring's HD-related stress were found to be significantly negatively associated with offspring's secondary control coping. *Discussion:* Results of the study can offer some understanding of factors that influence how offspring cope and can be further explored in the future in order to assess what interventions can be used to improve coping in offspring.

Introduction

Overview of Huntington's disease

Huntington's disease is an autosomal-dominant neurodegenerative disease that is caused by an expanded cytosine-adenine-guanine (CAG) repeat in the HTT gene, which codes the protein huntingtin (Huntington's Disease Collaborative Research Group, 1993). Due to the autosomal-dominant nature of the disease and complete penetrance of the expanded gene, offspring of a parent with Huntington's mutation have a 50% chance of inheriting the mutated gene, which will eventually result in development of the disease later in life (Huntington's Disease Collaborative Research Group, 1993). In carriers of the mutated gene, the mutation causes the protein huntingtin to be produced in a way that predisposes the protein to fragmentation (Bates et al., 2015). The average age of onset of motor symptoms for Huntington's disease is 40 years old and the duration of the illness is around 15-20 years (Snowden, 2017). Due to the neurodegenerative nature of the disease, symptoms progressively worsen with time until death (Snowden, 2017).

Symptoms of the disease include a wide range of motor, cognitive, and neuropsychiatric impairments. Some examples of common motor impairments in patients with Huntington's disease include chorea and bradykinesia (Snowden, 2017). Chorea involves rapid involuntary movements of face and limbs and bradykinesia involves the slowing down of movement execution, and these symptoms can cause numerous challenges in daily life. Some examples of common cognitive impairments in patients with Huntington's disease include difficulties in planning and organizing, in multitasking, in memory (Cleret de Langavant et al., 2013; Snowden, 2017), in processing emotion (Sprengelmeyer et al., 1996; Snowden, 2017), in speech production

(Podoll et al., 1988; Snowden, 2017), and with attentional and inhibitory control (McDonnell et al., 2020). The reduced ability to plan ahead and organize can lead to a decreased ability to complete simple daily tasks. The loss of the ability to efficiently multitask can also cause a Huntington's disease patient to have reduced ability to efficiently accomplish tasks like they used to, which can especially be hard in a workplace setting. Huntington's disease can often interfere with the HD-diagnosed patient's ability to recall information freely and it can also interfere with the patient's procedural memory, or the ability to learn things. In addition to motor and cognitive symptoms, neuropsychiatric symptoms are also prevalent in patients diagnosed with Huntington's disease. Some examples of common neuropsychiatric symptoms in patients with Huntington's disease include irritability, apathy, anxiety, and depression (Craufurd & Snowden 2014; Snowden, 2017). Apathy involves the loss of motivation to complete tasks and this can significantly impair a HD-diagnosed patient's ability to function.

The Burden of Huntington's Disease

Huntington's disease is not only burdensome because of the physical and cognitive impairments, but it is also very burdensome due to the stress associated with the diagnosis and the symptoms of the illness (Craufurd & Snowden, 2014; Snowden, 2017). Huntington's disease is not only stressful to patients but it is also stressful to family members, caregivers, and loved ones of patients (Downing et al., 2011; Jona et al., 2017; Roscoe et al., 2009). Huntington's disease can be highly stressful for patients for various reasons. For example, because of the genetic nature of the disease, Huntington's disease diagnosed patients likely witnessed one of their parents suffer from the disease and the effect it had on their parents and this can cause stress due to fear of experiencing the same (Helder et al., 2002; Tyler, 1991). Patients also face

enormous amounts of stress due to the neurodegenerative nature of the disease which can worsen motor and cognitive capacities over time, and can cause great uncertainty about the future. Patients diagnosed with Huntington's disease also often face social stigma, unemployment, economic dependency, and physical dependency on others, causing increasing burdens on someone already faced with the stress of worsening symptoms (Hayden, Ehrlich, Parker & Ferera,1980; Helder et al., 2002; Tyler, 1991). Patients can also experience stress due to feeling like a burden to others and due to feeling responsible for potentially passing down the disease to their offspring (Helder et al., 2002). Additionally, pre-symptomatic genetic testing, following the discovery of the mutated gene related with Huntington's disease, has made it possible for patients to get diagnosed long before they experience or manifest any symptoms (Helder et al., 2002), which can cause lots of stress for those diagnosed who know their life will change drastically in the future.

Family caregivers and offspring of a parent with Huntington's disease can also face high levels of stress from the disease as well for various reasons. Younger offspring will likely face significant stress due to the uncertainty of whether they will get the disease or not and adult offspring who have tested positive can often face stress about their own future in dealing with the disease. Adult offspring caregivers will likely face additional stress of having to take care of their parents and seeing their own parent's situation getting worse, while dealing with the potential diagnosis of the disease themselves in the future (Roscoe et al., 2009). Even if the adult offspring caregivers have tested negative for Huntington's Disease, they will still likely face a significant level of stress from seeing their relationship with their parents deteriorate and for having to spend their own adult years taking care of their diagnosed parent. The adult offspring caregiver

may also face significant stress from neglecting their own parental duties to help their own parents if they have offspring of their own, and they may suffer from uncertainty about passing the disease on to their own offspring. Spousal caregivers likely will face high levels of stress dealing with and having to take care of their own spouse while also worrying about if their offspring inherit the disease, and whether they will have to take the responsibility of taking care of both a spouse and a son or daughter with Huntington's disease (Roscoe et al., 2009). Overall, Huntington's disease diagnosis is very stressful for all members of the family and this stress can be very hard alongside the illness itself.

Coping with Huntington's Disease Related Stress

Because Huntington's disease can cause high levels of stress in patients and their families, adaptive coping skills can be important for managing the severe amounts of stress that comes with Huntington's disease. Coping with stress involves three primary voluntary factors, which include primary control engagement coping, secondary control engagement coping, and disengagement coping (Compas et al., 2001, 2017). Primary control engagement coping involves coping mechanisms involved in taking action to reduce the stressor itself or in taking action to change the emotions related to stressors. Primary control coping consists of mechanisms, such as problem solving, emotional expression, and emotional regulation/modulation. Problem solving involves doing something to fix a stressful situation to get rid of a stressor and this is helpful in situations where a stressor is controllable as problem solving will help reduce the stressor itself (Compas et al., 2001, 2017). Emotional expression is freely conveying and expressing emotions about stressors (Compas et al., 2001, 2017), and this can be helpful, because it involves addressing the emotions rather than suppressing them. Emotional regulation/modulation involves

taking action to reduce emotions (regulation) or dealing with emotions at an appropriate time (modulation) and this can be very useful, because it involves taking steps to change emotional reactions to stressors (Compas et al., 2006; Compas et al., 2014).

Secondary control coping usually involves coping mechanisms used in response to severe stressors that cannot be directly acted upon or changed (such as symptoms of a major illness). Examples of secondary control coping mechanisms include distraction, acceptance, positive thinking, and cognitive restructuring (Compas et al.,2001, 2017). Distraction involves voluntarily thinking of something other than the stressor (Compas et al., 2001, 2017), and this can be useful, because it can help to diminish negative feelings associated with ruminating about the stressor. Acceptance involves learning to accept the situation for what it is instead of trying to change it (Compas et al., 2006), and this can be useful, because it involves accepting the uncontrollable stressor for what it is. Positive thinking involves thinking about the positives of the stressful situation rather than the negatives (Compas et al., 2001, 2017), and this can be helpful, because it involves voluntarily viewing an undesirable situation in a positive light. Cognitive restructuring involves changing one's mindset from negative to more positive overall so that each uncontrollable stressor one faces can be thought of in a positive way (Compas et al., 2001, 2017), and this is similar to positive thinking in that both involve voluntary efforts to think about uncontrollable stressors in more positive lights.

Disengagement coping usually involves coping mechanisms that are not useful in managing symptoms of stress and can even make the stress worse. Disengagement coping mechanisms include avoidance, denial, and wishful thinking (Compas et al., 2001, 2017). Avoidance involves trying to purposefully forget about the stressor, and this can be detrimental

because it can cause a person experiencing stress to think about the stressor more. Denial involves denying the severity or reality of a stressful situation, and this can be detrimental, because denying that the stressor can lead to a build-up of suppressed emotions, which will probably make the stress even worse in the long run. Wishful thinking involves thinking that a stressor will just go away or resolve on its own and this will likely make things worse in the long run if the stressor does not resolve or go away and will likely lead to a build-up of stress and emotions.

A small number of prior studies have examined different aspects of coping with Huntington's Disease (HD)-related stress. For example, Helder et al. (2002) investigated the relationship between patients' illness perceptions, coping mechanisms, and well-being and found that illness perceptions and coping mechanisms are significant predictors of well-being of a patients, and overall the study highlights the importance of coping with HD-related stress (Helder et al., 2002). Additionally, Roscoe et al. (2009) looked at family caregivers of patients with late-stage Huntington's disease and they examined the relationship between positive appraisals of the caregiving experience and life satisfaction and health (Roscoe et al., 2009). Overall, Roscoe et al. (2009) found that the usage of positive appraisals (which is an example of a secondary control coping mechanism) was significantly correlated with better life satisfaction and health (Roscoe et al., 2009). More recently, Ciriegio et al. (2020) investigated the relationship between secondary control coping and symptoms of anxiety and depression in offspring of patients with Huntington's Disease, and they found that offspring's secondary control coping was significantly negatively correlated with symptoms of anxiety and depression.

Overall, these studies showed the importance of beneficial coping strategies in families with a HD-diagnosed member.

Goals for Honors Project

Despite the existing literature that involves examining the positive effects of coping strategies in family members and patients diagnosed with Huntington's disease, until recently, few studies have examined how offspring in particular cope with HD-related stress. Also, no studies, to my knowledge, have examined what factors may affect different types of coping with HD-related stress in offspring. I believe that because it is clear that secondary control coping is beneficial in reducing symptoms of anxiety and depression and improving quality of life, the next step would be to examine what accounts for differences in different subtypes of coping (primary control coping, secondary control coping, and disengagement coping) in offspring of parents with Huntington's disease. I believe it is very important to analyze the factors that affect differences in coping in the offspring, because understanding the factors that affect offspring's ability to use effective coping strategies could help to show which groups of offspring are more at risk of developing psychiatric disorders. If there are significant relationships found between several different variables and offspring's coping, then certain offspring could potentially benefit more from interventions in learning coping skills than others.

Research Question/Hypotheses:

For this study I plan on investigating (1) how age is related to reported levels of primary control coping, secondary control coping, and disengagement coping with HD-related stress in offspring; (2) how measures of disease progression in parents (number of CAG repeats) are directly related to use of primary control coping, secondary control coping and disengagement

coping in offspring; (3) do parents and their offspring cope similarly; and (4) if there is an indirect effect of disease progression in parents on how offspring cope with HD-related stress through perceived levels of offspring HD-related stress.

For this study, I propose to test several hypotheses, which include: (1) age of offspring would be significantly positively associated with secondary control coping in offspring of parents with Huntington's Disease, and have no significant association with primary control coping or disengagement coping; (2) CAP (CAG X age product) score in parents & number of CAG repeats will be significantly negatively associated with secondary control coping but will show no significant association with primary control coping or disengagement coping; (3) levels of perceived Huntington's Disease (HD) stress for offspring will be negatively associated with primary and secondary control coping and significantly positively associated with disengagement coping in the offspring; (4) parents' levels of reported primary control coping will be significantly positively correlated with offspring's primary control coping but show no significant association with offspring's secondary control coping or disengagement coping; (5) parents' levels of secondary control coping will be significantly positively correlated with offspring's secondary control coping but show no significant association with offspring's secondary control coping or disengagement coping; (6) parents' levels of disengagement coping will be significantly positively correlated with offspring's disengagement coping and will be significantly negatively correlated with offspring's secondary control coping and primary control coping; and (7) there will be an indirect association between parents' CAP score and secondary control coping in the offspring through levels of perceived offspring's HD related stress such that

CAP score predicts the offspring's ability to use less secondary control coping through the offspring's levels of perceived HD-related stress.

Method

Participants

This study includes data from 76 offspring of parents with Huntington's disease. The offspring in this study includes a mixture of both adolescent (ages 10-19) offspring and young adult (ages 20-39) offspring. All offspring in the sample have parents who are aged 30 or older and who have been diagnosed with Huntington's Disease. Eligibility requirements for these participants include: (a) English must be the participants' primary language, (b) parents of offspring must have the genetic mutation for Huntington's, (c) offspring's parents must be over 30 years of age, (d) offspring must be either adolescents (10-19) or young adults (20-39), and (e) parents with HD may range in disease severity (i.e., pre-symptomatic, prodromal, manifest) (see Ciriegio et al., 2020).

Measures

Coping

The Responses to Stress Questionnaire–Huntington's Disease Version (RSQ-HD) is a self-report measure that has been administered to all the offspring and parents to identify coping strategies in response to stress from being part of a family that is affected by Huntington's Disease (Ciriegio et al.,2020). The RSQ-HD has also been administered to the parents and offspring to measure perceived levels of HD-related stress. To measure coping, the second portion of the RSQ-HD was used. It includes 57 items reflecting coping (both automatic and voluntary responses) to stressors, where each item is rated on a 4-point scale (1 indicating not at

all and 4 indicating a lot). Examples of items on this portion include: “I think about the things I’m learning from being from a family affected by HD, or that something good will come from it” and “I tell myself that I can get through this, or I will be okay.” After all this, proportion scores were computed to identify three factors used in this study: primary control coping, secondary control coping, and disengagement coping.

To measure parents’ self-reported perceived HD stress, the first portion of the RSQ-HD patient edition was used, and this portion contains a list of ten representative stressors. The patients rank how stressful various HD-related stressors are using the 4-point scale mentioned earlier. Example items of this include: “Progressive loss of communication skills” and “concerns about my future.”

Huntington’s Disease Progression

Disease severity is looked at using CAP (CAG-Age Product) Scores and by number of CAG repeats in the parents. In order to measure CAP scores, CAG repeat length is needed. As mentioned earlier, a mutation in the HTT gene that causes an abnormally large CAG repeat length is what causes Huntington’s disease. This repeat length varies among patients and has an effect on disease severity, due to the involvement in the production of the protein, Huntingtin. Parent CAP scores have already been calculated for all participants using this equation: (CAG repeat length -33.66) x age (Zhang et al., 2011) and these scores measure disease progression. Higher CAP scores indicate more severe disease progression. Another indicator of disease severity includes parent’s observed chorea. As mentioned earlier, chorea involves involuntary random movements. Parent’s observed chorea have been measured by a board-certified neurologist for all participants. The Unified Huntington's Disease Rating Scale total maximal

chorea score was calculated based on established criteria (Mestre et al., 2018). This score ranges from 0 to 28 with higher scores indicating greater chorea.

Design

This study is a cross-sectional study. The variables of the study include several parental factors: (1) reported primary control coping of the parents, (2) reported secondary control coping of the parents, (3) reported disengagement control coping of the parents, (4) parent's CAP score, and (5) number of CAG repeats in parents. The variables related to offspring will include: (1) reported primary control coping of the offspring, (2) reported secondary control coping of the offspring, and (3) reported disengagement control coping of the offspring, (4) age of offspring, and (5) offspring's perceived levels of HD-related stress.

Procedure

All the participants in this study were recruited through a Huntington Disease Society of America Level 1 Center of Excellence (COE) between October 2018 and December 2021. Informed consent and assent were obtained from all parents and offspring prior to study enrollment and participation. The medical director of the COE oversaw the recruitment of eligible families, and a member of the clinical team at the COE made the initial study introduction. This study is part of an ongoing investigation involving parents and offspring's ability to cope with Huntington's disease related stress, so informed consent and assent has already been obtained from all parents and offspring prior to study enrollment and participation. Following recruitment of participants, participants came into the Vanderbilt University Medical Center Department of Neurology between October 2018 and December 2021. During visits, both

parents and offspring filled out the RSQ-HD. In addition to this, parents also had their number of CAG repeats and CAP scores measured.

Statistical Analyses:

Means and standard deviations for all variables in this study were calculated using the Statistical Package for the Social Sciences (SPSS; version 28). Bivariate Correlations between CAG repeats, CAP scores, age of offspring, perceived levels of HD-related stress, primary control coping of offspring, secondary control coping of offspring, disengagement coping of offspring, primary control coping of parents, secondary control coping of parents, and disengagement coping of parents were calculated using the Pearson Correlation analyses function of SPSS version 28. To evaluate the indirect effect of CAP scores on secondary control coping through perceived levels of HD-related stress, Model 4 of the PROCESS Macro for SPSS was used.

Results:

Descriptive Statistics (means, standard deviations) for all the variables of the study are shown in Table 1. Bivariate Pearson Correlation analyses are reported in Table 2. These correlations reveal that no support for the first hypothesis was found. No significant correlations were found between age of offspring and usage of secondary control coping as measured by the RSQ-HD. Although the hypothesis was not supported, a significant negative correlation was found between age and disengagement coping on the RSQ-HD ($r = -.22$, $p < .05$), which reveals that there was a small association of age with disengagement coping. Despite no support for the first hypothesis, correlation analyses did support the second hypothesis as the CAP score was significantly negatively associated with secondary control coping levels in offspring ($r = -.30$, p

< .05). Additionally, the effect size of the correlations between CAP scores and secondary control coping was medium (r > .3). Number of CAG repeats was also significantly negatively associated with secondary control levels in offspring (r= -.25, p < .05) and was significantly positively associated with disengagement coping levels in offspring (r = .28, p < .05). In addition to the second hypothesis being supported, partial support for the third hypothesis was found as perceived levels of HD-related stress measured by the RSQ-HD were found to be significantly positively correlated with secondary control coping (r = -.74, p < .001). In addition, the effect size of the correlation was large (r > .5). Despite this finding, no significant correlations were found between perceived levels of stress and measures of primary and disengagement coping in the offspring.

The Pearson correlations also revealed full support for the fourth hypothesis. Parents' levels of primary control coping as measured by the RSQ-HD was significantly positively correlated with offspring's primary control coping levels (r = .30, p <.05). The effect size of this correlation was small (r < .3). In addition to this, parents' primary control coping had no significant association with the offspring's secondary control or disengagement coping, which is consistent with the hypothesis. Despite parents' primary control coping being related with the offspring's primary control coping, no support was found for the fifth hypothesis. Parents' secondary control coping as measured by the RSQ-HD showed no significant correlation with the offspring's usage of secondary control coping. Despite no support for this hypothesis, partial support was found for the sixth hypothesis. Parent's disengagement coping as measured by the RSQ-HD was significantly negatively correlated with the offspring's usage of primary control

coping. Despite this finding, no significant associations were found between parents' disengagement coping and offspring's secondary control and disengagement coping.

Results of the mediation analysis using PROCESS to test the indirect effect between parents' CAP scores and secondary control coping through perceived levels of HD-related stress is shown in Figure 1. This analysis failed to support the seventh hypothesis, as no indirect association was found between CAP scores and secondary control coping in the offspring through perceived levels of HD-related stress. The *a* path of the model (direct association of CAG repeats and perceived levels of stress) was not significant (β = .1641, p > .05). The b path of the model (direct association between perceived levels of stress and secondary control coping) was found to be significant (β = -.67, p <.01). The c path of the model (direct association between CAG repeats and secondary control coping) was not found to be significant (β = -.36, p > .05). The c' path of the model (direct effect of CAG repeats on secondary control coping) was not found to be significant as well (β = -.25, p > .05). The indirect effect of CAP scores on secondary control coping through perceived levels of HD-related stress (ab path) was also not found to be significant (β = -.11, SE = .18, 95% CI [-.52,.21].

Discussion

This study examined several potential factors that could be associated with and influence how offspring of parents diagnosed with Huntington's Disease cope with HD-related stress. As mentioned earlier, offspring of parents diagnosed with Huntington's Disease face many stressors as a result of their parent's diagnoses with the disease and as a result of their uncertainty about their own diagnosis (Roscoe et al., 2009). For this reason, it is not only important that offspring use higher levels of primary control and secondary control coping and low levels of

disengagement coping, but also to examine what factors influence the offspring's ability to successfully cope with the stress.

With regard to the first hypothesis that was tested, age was found to have no significant association with secondary control coping. This was inconsistent with the hypothesis that age would be significantly positively associated with secondary control coping. The Pearson correlation coefficient ($r = -.051$) for this relationship is almost 0, so this finding indicates that age seems to be unrelated to offspring's usage of secondary control coping. Age was, however, shown to be significantly negatively associated with disengagement coping. Although the effect size was small, this finding does indicate that older offspring may use less disengagement coping than younger offspring. Although this finding is significant, the lack of relationship between age and secondary control coping indicates that overall age is probably not a useful predictor of how offspring of HD-diagnosed parents cope with stress. These findings are overall inconsistent with previous literature that showed associations between age and coping (e.g., Nolen-Hoeksema & Aldao, 2011).

In support of the second hypothesis, the patients' CAP score was found to be significantly negatively associated with secondary control coping, and no relationship was found between CAP score and primary control or disengagement coping. Additionally, number of CAG repeats was also found to be significantly negatively associated with secondary control coping and was found to be significantly positively associated with disengagement coping. CAP scores are one of the most important measures of Huntington's-Disease severity, and so this finding indicates that offspring of parents with greater disease severity tend to use less secondary control coping. Additionally, CAG repeats were found to be significantly positively associated with

disengagement coping, which indicates that higher CAG repeats in parents is associated with higher disengagement coping strategies, As mentioned earlier, secondary control coping strategies are coping strategies used when dealing with uncontrollable stressors, such as those directly related to the disease, and disengagement coping are harmful coping strategies that do not adequately alleviate stress levels. Knowing that one marker of disease severity (CAP score) is related to secondary control coping is important because it indicates that usage of these strategies could be hindered by how severe their parent's disease is. Additionally, knowing that another measure of disease severity (number of CAG repeats) is associated with disengagement coping is important because it indicates that higher disease severity can lead to more usage of harmful coping strategies.

In partial support of the third hypothesis, perceived levels of HD-related stress were found to be significantly negatively correlated with secondary control coping. No significant correlations were found between perceived levels of HD-related stress and primary control or disengagement coping. The correlation was not only significant but also was very strong. These findings suggest that higher levels of perceived stress caused by the disease is associated with offspring using less secondary control coping strategies. These findings are important, because they indicate that greater levels of perceived stress arising from the disease could impact the offspring's abilities to successfully cope with stress arising from Huntington's Disease.

In full support of the fourth hypothesis, parents' reported levels of primary control coping were found to be significantly positively associated with offspring's levels of primary control coping. In addition, parent's levels of primary control coping were found to have no significant association with offspring's levels of secondary control coping. Parents' use of primary control

coping strategies seemed to influence the offspring's use of primary control coping strategies. Given that primary control coping strategies are especially useful when dealing with daily stressors that can be controlled, it is important to know that how the parents use these strategies seems to slightly influence how the children use these strategies.

Although support of the fourth hypothesis was found, there was no support found for the fifth hypothesis. HD-diagnosed parents' reported secondary control coping had no significant correlation with how the offspring coped with HD-related stress. HD-diagnosed parents' secondary control coping also had no correlation with offspring's primary control or disengagement coping. Given that parents' primary control coping was significantly correlated with the offspring's primary control coping, it is surprising that parents' secondary control coping had no significant correlation with the offspring's secondary control coping strategies. Although this result did not support the hypothesis, it is important to know that the usage of secondary control coping strategies in HD-diagnosed parents does not correlate with the usage of secondary control coping strategies in offspring.

In partial support for the sixth hypothesis, disengagement coping was found to be significantly negatively correlated with the offspring's primary control coping. This result indicates that higher use of disengagement coping in parents is associated with less use of primary control coping strategies in the offspring. Despite this finding, disengagement coping in parents was not found to be significantly correlated with the offspring's disengagement or secondary control coping.

Finally, no support for the seventh hypothesis was found. There was no indirect effect of CAG repeats on offspring's secondary control coping through offspring's perceived levels of

HD-related stress. The mediation analysis clearly showed that stress did not mediate the relationship between CAG repeats and secondary control coping.

Overall, the findings of this study failed to completely support all of the hypotheses, but partial support was found for many hypotheses. The most significant findings were that of CAG repeats and its association with secondary control coping in offspring, CAG repeats and its association with disengagement coping, the associations between perceived levels of stress and secondary control coping, and the associations between parents' primary and disengagement coping with offspring's primary control coping. Although most of the proposed hypotheses were not fully supported by the analyses, the results showed some very important information that can be expanded on in the future in order to accurately discover the best approach to improve primary control and secondary control coping and decrease disengagement coping in offspring of parents diagnosed with HD. Finding no correlations between age and secondary or primary control coping and finding a significant negative correlation between age and disengagement coping was a useful finding, because it indicated that overall age of offspring did not influence how they cope overall, which means that any future interventions to improve coping can be targeted towards all age groups. The finding involving older age of offspring being related with less usage of disengagement coping had a small effect size, and could be investigated further in the future using a longitudinal design to see if disengagement coping does decrease with age naturally. Overall, age was not a major factor in this sample's ability to cope with HD-related stress, so there could be something about the disorder itself that causes this relationship that has been found between age and coping in other populations to not exist.

The lack of an association between parents' secondary control coping and the offspring's secondary control coping was also important to look at and the lack of associations between parent's disengagement coping and the offspring's disengagement coping are important, because it showed that some difference exists between how the parents cope and how their offspring copes with Huntington's Disease-related stress. Future studies could look at potential reasons for why this difference exists in this population, and could implement tools to see what can be done to improve the coping strategies of all the members of a family affected by Huntington's Disease. The finding that parent's primary control coping was associated with the offspring's primary control coping was important as well, because it indicated that parent's usage of primary control coping may influence how their child copes with stress, and if parents do not use effective primary control coping methods, their offspring could be at risk for not using these methods as well.

The finding that parents' CAG repeat scores, number of CAG repeats, and perceived levels of HD-related stress were all significantly negatively correlated with secondary control coping was important, because they show factors that are associated with less usage of secondary control coping in offspring. Unfortunately, the mediation analysis did not find an indirect association between CAG repeat scores and secondary control coping through perceived levels of HD related stress, but the correlations still highlight important factors that appear to influence offspring's secondary control coping. Future studies could expand upon the CAP score and CAG repeats findings and could measure how many factors of disease progression influence how offspring cope, in order to see if how bad the disease is in parents significantly decreases secondary control coping in the offspring. This future research could then help develop potential

intervention programs to help offspring of parents with severe Huntington's Disease cope with the stressors better.

Strengths of the study design included the usage of validated measures to measure primary control coping, secondary control coping and disengagement coping (RSQ-HD) as well as to measure the perceived levels of stress. The measures of disease severity in the form of CAP score and CAG repeats were also a strength of the study, as this score reflects Huntington's Disease severity very well. Several limitations of the study could have influenced results. For example, the small sample size of the study could have influenced the power to test the correlations. With larger sample sizes, there could have been more significant correlations that were found that could then be able to better explain the factors that influence coping. Another limitation is that only one aspect of disease progression (CAP score and CAG repeats) was looked at. If more measures of disease progression are looked at, it could more accurately show if the parent's disease progression does have an influence on secondary control coping in the offspring. Another limitation is that among the few findings of significant correlations, most correlations either had small or medium effect sizes. Although findings are significant, the lack of a large effect size means that many of the relationships between factors and different types of coping are likely on the smaller end. Calculating Pearson correlations in the future with more participant data could be useful to see if these correlations on the smaller end are found again with a larger sample. Finally, the biggest limitation of the study is probably the lack of significant correlations found between the variables and different types of coping. In the future, even more variables can possibly be looked at, and more mediation analyses can be run in order to find the variables that have the biggest influence on coping. Looking at the factors that

influence coping in offspring of HD-diagnosed parents is very important and should continue to be researched. Once more research is done on the factors that influence coping, different intervention programs can then be tested to see how to help improve useful coping strategies and decrease negative coping strategies in this population.

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Supplementary Materials:

Table 1

Descriptive Statistics (Range, Mean, and Standard Deviation) for measures of the study

Constructs	Range	Mean	Standard Deviation
Age	7-38	20.85	8.15
Sf36 physical functioning	0-100	62	29.12
Sf36: emotional well-being	8-96	63.7	21.27
Offspring Primary control coping	.08-.28	.17	.04
Offspring Secondary control coping	.12-.41	.27	.06
Offspring Disengagement coping	.09-.25	.15	.03
Perceived levels of HD-stress	10-34	18.35	6.62
CAP score	251.72- 702.62	448.07	101.91
Number of CAG repeats	38-50	43.66	2.90
Parent Primary Control Coping	.11- .26	.18	.04
Parent Secondary Control Coping	.11-.39	.26	.06
Parent Disengagement Coping	.07-.21	.14	.03

Table 2

Pearson Bivariate Correlations between all variables of the study

Constructs	1	2	3	4	5	6	7	8	9
1. Age of offspring	-								
2. Parent's Primary control coping	.06	-							
3. Parent's secondary control coping	-.11	.37**	-						
4. Parent's disengagement coping	-.00	-.69**	-.38**	-					
5. Levels of Perceived HD stress	-.10	-.70**	-.69**	.70**	-				
6. CAP score	.01	.06	-.36**	.10	.17	-			
7. # of CAG repeats	-.52**	.05	-.25	.14	.15	.75**	-		
8.Offspring's primary control coping	.14	.30*	.07	-.30*	-.25	.06	-.19	-	
9.Offspring's secondary control coping	-.05	-.05	.15	-.02	-.74**	-.30*	-.25*	.24*	-
10.Offspring's Disengagement coping	-.22*	-.06	.09	.19	-.10	.10	.28*	-.47**	-.30**

*Note. * represents correlation is significant at the .05 level (2- tail).*

*** represents that correlation is significant at the .01 level (2-tail).*

Figure 1.

Results of the Mediation Analysis that tested if there was an indirect effect of CAG repeats on offspring's secondary control coping through perceived levels of offspring's HD-related stress.