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Influence of Parent Chronic Pain History on Youth's Experience of Hypermobility Ehlers-Danlos Syndrome

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Influence of Parent Chronic Pain History on
Youth's Experience of Hypermobile Ehlers-Danlos Syndrome

A Thesis

Presented in

Partial Fulfillment of the
Requirements for the Degree of
Master of Arts

By

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Abstract

The current study examines parent factors that may relate to youth's experiences with Hypermobile Ehlers-Danlos Syndrome (hEDS). hEDS, its symptomology, and associated psychosocial and physical outcomes are reviewed. A model of transgenerational transmission of risk associated with chronic pain is presented. Parents' own experiences with chronic pain is highlighted as an important determinant of how parents think about and respond to their child's pain. Potential pathways through which parent factors influence a child's own thinking about pain are investigated. The goal of the study was to learn more about parent factors that influence child pain-related outcomes and the pathways through which they exert their influence. It was hypothesized that parents with chronic pain or hEDS will be more likely to catastrophize about their child's pain and respond to their child's pain more protectively than parents without their own history of pain. It was additionally hypothesized that children of parents with chronic pain or hEDS would have worse psychosocial and functional outcomes than children and adolescents whose parent does not have chronic pain or hEDS. Effect sizes provided evidence for the opposite relationship in which children of parents with a positive pain history had better pain-related outcomes. A greater understanding of the transmission of risk within families affected by hEDS can inform future intervention and treatment for youth with hEDS to increase their efficacy and lead to more positive pain-related outcomes.

Introduction

Hypermobile Ehlers-Danlos Syndromes (hEDS) is a heritable connective tissue disorder characterized by joint instability and dislocations, chronic widespread joint pain and skin manifestations that affects about 255 million people worldwide (Mulvey et al., 2013; Tinkle et al., 2017). Disorder characteristics and common comorbid symptoms (e.g., chronic fatigue, headache, gastrointestinal dysfunction and urinary stress incontinence) negatively impact physical and psychosocial functioning (Pacey et al., 2015; Scheper et al., 2016; Tinkle et al., 2017). While symptom presentation varies considerably by individual, children and adolescents with hEDS are at risk for functional impairment, psychological distress (e.g., anxiety and depression) and reduced HRQoL across all domains (i.e. physical, emotional, social and school) (Engelbert et al., 2017; Fatoye et al., 2012; Mu et al., 2019). The literature on pediatric hEDS is limited compared to other chronic pain conditions, but it offers preliminary evidence that children with hEDS respond similarly to their pain experiences compared to youth with other chronic pain conditions, and so this literature is a valuable resource (Fatoye et al., 2012; Pacey et al., 2015).

An important contributing factor to youth's hEDS experience is that hEDS is inherited, and so, many children and adolescents with hEDS have a parent or other family members with hEDS (Castori et al., 2014). However, few aspects of parent-child relationships have been explored in the hEDS community (De Baets et al., 2017; Pacey et al., 2015). The relationship between parent and child chronic pain experiences and influence of parent-child interactions on child outcomes are gaps in the hEDS literature that need to be filled.

The current study seeks to help fill these gaps by examining patterns of disability and psychological functioning among pediatric hEDS patients in relation to parental chronic pain

history, parent pain-related cognitions, and parent driven pain-specific social learning as conceptualized by Stone and Wilson's Conceptual Model of Intergenerational Transmission of Chronic Pain Risk (Stone & Wilson, 2016). The model discusses multiple mechanisms by which parental chronic pain is a risk factor for the development of pediatric chronic pain (Stone & Wilson, 2016). These mechanisms are proposed to bidirectionally interact with child vulnerabilities, which in turn influence pain-related child outcomes such as chronic pain experience, disability, and psychological functioning (Stone & Wilson, 2016). While the model focuses on parents with a history of chronic pain, the proposed mechanisms interact with child vulnerabilities regardless of parental chronic pain history to influence child pain outcomes (Denk et al., 2014; Palermo & Chambers, 2005). However, past research leads us to believe that the interaction between mechanisms and child vulnerabilities may differ for children of parents with and without chronic pain (Palermo & Chambers, 2005; Wilson & Fales, 2015; Wilson et al., 2014) as parent's own chronic pain experiences, or lack thereof, inform mechanism pathways.

How a parent thinks about their own pain impacts and is impacted by their pain experiences and so, parents with chronic pain may think about pain and respond to pain differently than parents without chronic pain. Since a core symptom of hEDS is chronic pain, it is expected that parents with hEDS respond similarly to parents with other chronic pain conditions. Research suggests that parents with chronic pain may be more likely to catastrophize about their child's pain and respond to their child's pain with protective behaviors than parents without chronic pain (Langer et al., 2009; Wilson & Fales, 2015; Wilson et al., 2014). Perhaps parent's own experiences with pain make them more attuned to picking up on their child's pain cues, but also may increase their risk of catastrophizing about their child's pain and may increase the likelihood that they will engage in protective behaviors in response to their child's pain

(Wilson & Fales, 2015). In addition to teaching their children how to respond to pain through responses to their child's expressions of pain, parents also teach through modeling their illness behaviors and beliefs (Levy, 2010; Walker & Zeman, 1992). For instance, when parents stay at home from work or expect special privileges when they are in pain, they model these behaviors and expectations for their children. Similarly, attempts to make things easier for their child by allowing their child to skip chores, delay homework, and miss school, increase positive consequences of illness, promoting future pain expression (Levy, 2010). Rewarding children's symptomatic complaints also teaches the child to attend to their symptoms more and may lead them to become sensitized to picking up on lower thresholds of pain (Levy, 2010; Walker et al., 1991). Subsequently, these protective and solicitous parental behaviors are related to increased pain and disability in children with chronic pain (Claar et al., 2008; Langer et al., 2009; Levy, 2010).

With regard to child vulnerabilities, how a child thinks about their own pain influences their pain experience. Children who engage in more pain catastrophizing are at risk for greater pain intensity, depressive symptoms, anxiety symptoms, functional disability, and reduced quality of life (Langer et al., 2009; Lynch-Jordan et al., 2013; Pielech et al., 2014). Due to negative outcomes associated with higher pain-catastrophizing, understanding how children come to develop catastrophizing beliefs is vital. Parents play a role in influencing child pain catastrophizing (Cunningham et al., 2014; Pielech et al., 2014; Welkom et al., 2013). Thus by influencing child pain beliefs, they subsequently influence their child's psychosocial and functional outcomes (Lynch-Jordan et al., 2013; Pielech et al., 2014; Wilson et al., 2014). Parental influence has both genetic (Troost et al., 2015) and social learning roots (i.e. protective response behaviors (Langer et al., 2009; Wilson & Fales, 2015)). For example, parents who

exhibit a greater frequency of protective parenting responses tend to have children with stronger pain catastrophizing beliefs, which in turn, predicts increased functional disability in youth (Cunningham et al., 2014; Welkom et al., 2013).

When considered altogether, studies suggest that in response to observing their child in pain, parents who exhibit higher levels of pain catastrophizing, respond with more protective behaviors. Protective behaviors teach and reinforce the child's catastrophic thinking about pain, which reduces effective coping with pain, leading to increased child pain intensity, functional disability, and psychological distress. Because it has been suggested that parents with chronic pain may be more likely to catastrophize about pain and pass these beliefs onto their children through increased use of protective response behaviors, parents with hEDS may put their children at additional risk for increased pain intensity, functional disability, anxiety, and depression, relative to parents without a history of hEDS or other chronic pain condition. It is clinically important to identify risks for worse pain-related child outcomes and the pathway through which they are transmitted in order to design and effectively implement targeted family treatment and prevention programs that minimize risks and seek to improve HRQoL for children with hEDS.

These relationships have yet to be explored within the pediatric hEDS community, thus the current study aims to examine these parent-child relationships. Our first hypothesis is that there will be a positive association between parent pain catastrophizing beliefs, protective parental response behaviors, child pain catastrophizing beliefs, pain intensity, functional disability, anxiety symptoms, and depressive symptoms in children with hEDS. Second, we hypothesize that parents with hEDS or other chronic pain will have higher pain catastrophizing and report more protective response behaviors than parents with no history of chronic pain.

Third, we hypothesize that children of parents with hEDS or other chronic pain will have worse pain-related outcomes (i.e., pain intensity, functional disability, symptoms of anxiety and depression), than children of parents without hEDS or chronic pain. Fourth, we hypothesize that parental pain catastrophizing will predict child-pain related outcomes, through the effect that parent pain catastrophizing has on protective parental response behaviors. Fifth, we hypothesize that parental pain catastrophizing will predict child-pain related outcomes through the effect that parent pain catastrophizing has on child pain catastrophizing. Finally, we hypothesize that parental protective response behaviors will predict child-pain related outcomes through the effect that parental protective response behaviors have on child pain catastrophizing.

Method

Participants

Youth with hEDS and their parents were recruited in person during an hEDS clinic appointment within the clinical genetics division at a Midwestern children's hospital. Patients were screened for eligibility by the study geneticist during their medical appointments. Patients who were diagnosed with hEDS using the Villefranche criteria (Beighton et al., 1998) were eligible to participate. Inclusion criteria additionally required youths to be between 8 and 18 years old, speak and read English fluently and be cognitively able to assent and answer study questionnaires.

Procedures

Clinic families who provided consent and assent were provided parent and child questionnaire packets to complete either in clinic or to mail back. Both the Institutional Review Board (IRB) at the participating Midwestern university and children's hospital provided study approval.

Measures

Demographic information. The following demographic information was collected: patient and caregiver age, sex, race, and ethnicity; caregiver relationship to patient, family income, insurance type, and time since patient hEDS diagnosis.

Parent Measures

Parental Pain History. Parents reported family history of hEDS and chronic pain. Their own pain status was categorized as either positive pain history (e.g. have hEDS or other chronic pain condition) or no pain history (e.g. do not have hEDS or other chronic pain condition).

Pain Catastrophizing Scale Parent Version (PCS-P). The PCS-P is a 13-item parent self-report measure that assesses parent catastrophic thinking about their child's pain on a 5-point scale (0 = *not at all*, 4 = *extremely*) (Goubert et al., 2006). Items include "When my child is in pain..." "I keep thinking about how much I want the pain to stop," "I become afraid that the pain will get worse," and "it's awful and I feel that it takes over me." Scores range from 0 to 52 with higher scores indicating greater pain catastrophizing. PCS-P criterion validity and reliability were found in a sample of parents of adolescents with chronic pain (Goubert et al., 2006).

Adult Responses to Children's Symptoms (ARCS) Protect Subscale. The Protect subscale of the ARCS contains 13-items which assess how often a caregiver engages in behaviors in which the child receives special attention, treatment, privileges and reduced responsibility expectations in response to pain complaints using a 5-point Likert-type scale (0 = *never*, 4 = *always*) (Van Slyke & Walker, 2006). Example items include "When your child is in pain, how often do you..." "Stay home from work or come home early (or stay home instead of going out or running errands)," and "Tell your child that he/she doesn't have to finish his/her homework." Scores range from 0 to 52 with higher scores indicating greater use of protective

response behaviors. This structure of the ARCS Protect Subscale is suggested for use with combined child and adolescent populations and has been validated and strong reliability found in a sample of multiple pediatric chronic pain conditions and pain-related illnesses (Noel et al., 2015).

Child Measures

Pain Catastrophizing Scale Child Version (PCS-C). The PCS-C is a 13-item self-report measure that assesses children and adolescent's catastrophic beliefs about their own pain experiences (Crombez et al., 2003). It parallels the PCS-P, but has the item prompt: "When I am in pain...". It uses the same 5-pt scale and scores range from 0 to 52 with higher scores indicating greater pain catastrophizing. The PCS-C has been validated for youth ages 8 to 16 with and without chronic pain (Crombez et al., 2003).

Pain Intensity. Children and adolescents reported their "usual level of pain in the last 2 weeks" on an 11-point numeric rating scale ranging from 0 = *No Pain at all* to 10 = *Worst Pain I Can Imagine*. This scale has been found to be a valid and reliable assessment of children's pain intensity (Castarlenas et al., 2017).

Patient Reported Outcomes Measurement Information System (PROMIS) Pediatric Anxiety Subscale. The Short Form Anxiety subscale contains 8 items that ask children to report how often they have experienced different anxious feelings over the past 7 days using a 5-point Likert-type scale, where 0 = *Never*, 4 = *Almost Always*. Sample items include, "I felt nervous," "I felt worried," and "I got scared really easy." Item responses are summed ranging from 0-32. Raw scores from the short-form measure are converted to scaled T-scores (mean = 50). Higher T-scores indicate more anxious symptoms. It is for use with children between the ages of 8 and 17

years living with chronic illnesses (Varni et al., 2014) and the short form has sufficiently provided a precise measure of symptoms (Irwin et al., 2010).

PROMIS Pediatric Depression Subscale. The Short Form Depression subscale contains 8 items and asks children to report how often they have experienced different depressive feelings over the past 7 days using a 5-point Likert-type scale, where 0 = *Never*, 4 = *Almost Always*. Sample items include, “I could not stop feeling sad,” “I felt lonely,” and “It was hard for me to have fun.” Item responses are summed ranging from 0-32. Raw scores from the short-form measure are converted to scaled T-scores (mean = 50). Higher T-scores indicate more depressive symptoms. It is for use with children between the ages of 8 and 17 years living with chronic illnesses (Varni et al., 2014) and the short form has sufficiently provided a precise measure of symptoms (Irwin et al., 2010).

Functional Disability Inventory (FDI). The FDI contains 15 items which measure “physical functioning and disability in youth with chronic pain” across home, school, recreational, and social domains (Kashikar-Zuck et al., 2011, p. 1) on a 5-point Likert-type scale (0 = *No Trouble*, 4 = *Impossible*). Youths are asked to rate how much “physical trouble or difficulty” they have doing activities including “Walking up stairs,” “Reading or doing homework,” and “Getting to sleep at night and staying asleep.” Item responses are summed to create a total score ranging from 0 to 60 with higher scores indicating greater pain-related disability. Disability level may be categorized as “No/Minimal Disability” ($FDI \leq 12$), “Moderate Disability” ($FDI 13 - 29$), or “Severely Disabled” ($FDI \geq 30$) (Kashikar-Zuck et al., 2011). The FDI has been widely used with youth between the ages of 8 and 18 years (Kashikar-Zuck et al., 2011). Strong internal consistency, test-retest reliability, and parent-child concordance have been reported (Claar & Walker, 2006).

Qualitative Responses. Parents and children responded to the open-ended question “What makes living with EDS easier?” Responses were thematically analyzed for coping themes. The first author, her research advisor, and a graduate and undergraduate research assistant each generated a list of codes for parent and child responses. The study team met to review codes, compared discrepancies and came to a group consensus on a final list of codes, which each team member then independently assigned to responses; multiple codes could be applied per response. The group met again to review code allocation and came to a group consensus on final response code(s). Example child codes include “my own understanding of hEDS,” “sleep/rest,” and “social support.” Parent codes included “having a diagnosis/ knowledge about the condition,” “exercise/staying active” and “quality medical care/support from medical professionals.” There were unique and overlapping codes across groups.

Data Analyses

Preliminary analyses were carried out using SPSS Statistics (Version 24), included whole sample descriptive statistics, and bivariate correlations to test Hypothesis I. The sample was then split by parent pain history group to examine group differences. Bivariate correlations were repeated within parent history groups. Main analyses included examination of differences in means of study variables across parent pain history groups (Hypothesis II and III) using t-tests and effect sizes. The latter were calculated in Microsoft Excel 2016 to further examine group differences with Hedge’s g correction factor for Cohen’s d , due to small sample size. Mediation analyses using PROCESS were planned to test Hypotheses IV-VI: the effect of parent pain catastrophizing on child outcomes, through the effects of parental protective response behaviors and child pain catastrophizing (Hypotheses IV-V) and the effect of parental protective response

behaviors on child outcomes through its influence on child pain catastrophizing (Hypothesis VI). However, these analyses were not conducted based on results of preliminary analyses.

To further analyze data representing influences on child outcomes, responses to the open-ended question “What makes living with EDS easier?” were examined. Analyses were guided by a transcendental or psychological phenomenological approach (Creswell & Poth, 2017) as well as by an inductive thematic analysis (Braun & Clarke, 2006). Patterns of semantic content across parent pain history groups were described via frequency and percentage of code usage and results were used to develop coping themes and implications for child pain-related outcomes (Braun & Clarke, 2006).

Results

The current study included data from 34 children and adolescents between the ages of 8 and 18 years ($M = 14.68$, $SD = 2.92$) and 28 of their parents or caregivers ($M = 46.50$, $SD = 7.58$). Descriptive statistics including percentages, means, and standard deviations are reported for demographic variables of interest in Table 1. Overall, participants were a majority female and White. Of the 34 children, 23.5% were male, 70.6% were female, and 5.9% did not report sex. Caregivers reported child’s race/ethnicity; 52.9% of youth were identified as non-Hispanic White, 20.6% as Hispanic, 8.8% as Biracial (5.9% Asian or Asian American and White, 2.9% as American Indian or Alaska Native and White) and 17.6% of child participants’ race and or ethnicity were not captured. Of the 28 caregivers, 89.3% were mothers, 7.1% were fathers and 3.6% were grandmothers. Caregivers identified themselves as 82.1% non-Hispanic White, 7.1% Hispanic, 7.1% Asian or Asian American, and 3.6% as Biracial (American Indian or Alaska Native and White).

Children's psychosocial outcomes were also examined. Nearly a third (32.4%) of youth reported moderate symptoms and 17.6% reported severe symptoms of anxiety. Similarly, 26.5% reported moderate symptoms and 20.6% reported severe symptoms of depression. In terms of functional disability, 52.9% of youth reported moderate functional disability and 20.6% reported severe functional disability.

Bivariate correlations were run to determine associations between parent and child variables of interest: protective parenting behaviors, parent and child pain catastrophizing beliefs, child pain intensity, child functional disability, and child symptoms of anxiety and depression (Hypothesis I). Positive correlations were found between pain intensity, functional disability, anxiety symptoms, depressive symptoms, and child pain catastrophizing. Parent variables were not associated with each other or any of the child variables. Results displayed in Table 2.

Split Sample Analyses: Parent pain history group demographics including child and parent age, sex and ethnicity and family income were examined for differences (Table 1). Age of the child and adolescent participants were found to be significantly different ($t = 2.13, p = 0.045$) such that youth in the no parent history group were significantly older than youth in the positive parent history group. There was also a trend towards significantly more boys in the positive parent history group. In regards to racial and ethnic distribution, both groups contained a majority non-Hispanic White participants. However, the percentage of Hispanic families in the positive parent pain history group was twice that of the no parent pain group. Yet, the no parent pain group represented greater racial diversity (e.g., families with Asian or Asian American and Biracial identities in addition to non-Hispanic White and Hispanic families).

Within the positive parent pain history group, the significant correlations found across the whole sample remained, with two exceptions. The relationship between child depressive symptoms and pain intensity dissipated ($r = 0.357, p = .175$) and a relationship between parent pain catastrophizing beliefs and child pain intensity ($r = .591, p = .02$) emerged. Within the no parent pain history group significant correlations were found only between depressive symptoms and child pain catastrophizing ($r = .578, p = .049$) and functional disability and anxiety symptoms ($r = .828, p = .001$).

Main Analyses

No significant differences in means were found between study variables across the two parent pain history groups using independent samples t-tests; however, small to medium effect sizes were found (Table 3). Small effect sizes were found for differences between parent pain catastrophizing beliefs ($g = 0.35$) and parental protective response behaviors ($g = 0.25$) such that parents without pain had higher pain catastrophizing beliefs and responded more protectively to their children than parents with a history of hEDS or chronic pain (Hypothesis II). Small to medium effect sizes were found for differences between child pain catastrophizing ($g = 0.54$), pain intensity ($g = 0.55$), anxiety ($g = 0.59$), depression ($g = 0.56$) and functional disability ($g = 0.39$), such that children of parents with no pain history exhibited worse pain-related outcomes than children of parents with a history of hEDS or chronic pain (Hypothesis III).

Due to lack of association between parent and child variables, mediation analyses to examine the effect of parent pain catastrophizing on child outcomes, through the effects of parental protective response behaviors and child pain catastrophizing were not conducted (Hypotheses IV-V). Nor was the effect of parental protective response behaviors on child

outcomes through its influence on child pain catastrophizing (Hypothesis VI) as associations between these variables were not found.

Sequential Analyses

Quantitative analyses demonstrated that the directionality of the relationship between parent pain history and child outcomes was contradictory to prediction. As such, the researchers decided to present qualitative data that helps to increase understanding of the quantitative results. Parent and child responses to the question “What makes living with EDS easier?” were examined, and the number and percentage of coping themes were compared across parent pain history groups (Table 4). There were nine children and adolescents and ten parents in the no parent pain history group who provided responses and 13 youths and 12 parents in the positive parent pain history group. Twenty-two percent of children of parents without hEDS or chronic pain endorsed that their own understanding of hEDS makes living with hEDS easier, while no youth in the positive parent pain group did. More children of parents who do have hEDS or chronic pain indicated that activity pacing or setting limits (15.4% vs 0%), exercise or staying active (28.6% vs 11.1%), and medication (30.8% vs 22.2%) made life easier for them. Both sets of children and adolescents discussed positive physical attributes of having hEDS, others understanding of hEDS, social support, sleep or rest, distraction or keeping busy, physical therapy, and complementary or integrative techniques as helpful coping strategies.

More parents without hEDS or chronic pain indicated that having a diagnosis or knowledge about the condition (40% vs 8.3%) and school accommodations (30% vs 0%) help make living with hEDS easier. Parents with hEDS or chronic pain uniquely discussed activity pacing or setting limits, sleep or rest, exercise or staying active and diet as helpful and more discussed medication use (25% vs 10%). Both sets of parents describe social support, quality

medical care and support from medical professionals, physical therapy or use of orthotics and braces, and complementary treatments as helpful coping strategies at similar rates.

Due to sex differences between parent pain history groups, pain-related outcomes and qualitative responses were analyzed for differences between boys and girls. The no parent pain history group only included one boy, so sex differences were not examined. Independent samples t-tests did not identify differences across child and parent outcomes by sex within the positive parent pain history group, however; small, medium and large effect sizes were found. A small effect ($g = 0.28$) was found between usual pain level such that girls ($M = 4.78, SD = 2.77$) expressed experiencing greater pain than boys ($M = 4.00, SD = 1.00$). A medium effect ($g = 0.48$) was found for youth pain catastrophizing such that boys ($M = 20.80, SD = 12.81$) engaged in more pain catastrophizing than girls ($M = 14.78, SD = 11.37$). A large effect ($g = 0.85$) was found for parent catastrophizing about their child's pain such that parents' catastrophized less about their son's pain ($M = 12.33, SD = 8.02$) than about their daughter's pain ($M = 21.38, SD = 11.21$). Qualitatively, within the positive parent pain history group, girls uniquely discussed positive physical attributes, other's understanding of hEDS, social support, distraction/keeping busy, while the boys uniquely discussed sleep or rest and exercise(s) or staying active as helpful coping strategies. This latter theme was identified by 57.1% of boys. Both boys and girls described medication, physical therapy and complementary and integrative techniques as helping to make living with hEDS easier.

Discussion

This study set out to examine whether there are differences between children's experiences with hEDS depending on their parent's pain history. We found that children and adolescents whose parent has a history of either hEDS or chronic pain experienced better

psychosocial and physical functioning than youths whose parent did not have a pain condition. We hypothesized about the pathways through which parental pain history exerts its influence on child outcomes but did not test those hypotheses quantitatively as preliminary analyses demonstrated that the proposed analyses were not appropriate. However, to further explore pathways through which parents influence their children's pain-related outcomes and to increase understanding of the unanticipated directionality of results that we found, we thematically analyzed open-ended qualitative responses. We suggest that the pathway through which parents exert an influence on their children's pain-related outcomes is informed by parent experience with pain and disease-related coping strategies. Specifically, children with parents with their own pain condition benefit from their parents' knowledge of effective disease management strategies and are able to enact these strategies to cope with their own pain and symptoms effectively leading to better psychosocial and physical functioning.

The directionality of our results run contrary to study hypotheses, and the extant pediatric chronic pain literature which links parental experiences of pain to negative child-pain related outcomes (Cordts et al., 2019). While previous research suggests that parents with chronic pain may be more likely to catastrophize about their child's pain and respond to their child's pain with protective behaviors than parents without chronic pain (De Baets et al., 2017; Wilson & Fales, 2015; Wilson et al., 2014), this was not the case in our sample. In research with mothers with hEDS, mothers discussed their "double role model" status and the struggle to balance wanting to protect their children (e.g., make their children's experience of pain more bearable) and wanting to ensure their child is able to cope with their hEDS and grow up to be independent (De Baets et al., 2017). For instance, there is an internal dilemma associated with going to work while in pain, when one's child cannot go to school due to pain (De Baets et al., 2017). On one hand, the parent

wants to be a positive role model and show her child what is possible despite having hEDS, but she also wants to stay home and take care of her child (De Baets et al., 2017). This internal dilemma equates to the decision to engage in a protective response behavior or not.

It is likely that parents in our sample have similar internal debates. Personal experience with or without pain likely shapes pain catastrophizing beliefs and contributes to the decision about how to respond to their child as well as what illness behaviors to model. Perhaps a relatively more limited understanding of hEDS among parents without a history of hEDS or chronic pain contributes to a greater tendency to catastrophize over their child's pain because there is more that is "unknown" about their child's pain. These parents are also not working from a "double role model" framework, and so, they may be more willing to respond protectively to their children's illness behaviors or may not perceive modeling illness behavior to carry as much weight as it is relevant less often.

The role that parent pain catastrophizing and parental response behaviors played in results was not as impactful as expected. We expected that these parent factors would significantly differ by parent pain history and that these differences would drive differences in child outcomes. However, group differences in these parent factors had small effect sizes, but a majority of the differences in child outcomes across parent pain history groups had moderate effect sizes. These discrepancies in strength of differences indicate that parent pain history is a significant determinant of child outcomes, but there are likely additional parent factor variables that significantly contribute to differences in youth psychosocial and physical functioning. Similarly, the lack of relationship found between parent factors and child outcomes may also be reflective of including too few relevant parent factors in the study as well as having a small sample size. As demonstrated in a mixed pediatric chronic pain sample, inclusion of parent

chronic pain features including pain frequency, number of pain locations, and pain intensity in addition to parent chronic pain status, as well as the inclusion of parent physical functioning (e.g., pain interference, physical functioning), and additional parent psychological factors (e.g., anxiety, depression), in addition to parental pain catastrophizing, are needed to provide a thorough account of parental influence on child pain experience (Cordts et al., 2019).

Another potential important parent factor is knowledge of effective hEDS management strategies. Interpretation of qualitative responses from parents and children about what makes living with hEDS easier, indicate that parents with hEDS or chronic pain may be better informed about what active coping strategies will help with their child's illness management and improve mental health. We believe additional knowledge likely stems from personal experience with trial and error and additional years working with specialists. For instance, parents with hEDS or chronic pain uniquely discussed activity pacing and limit setting (e.g., one parent wrote "He...has limited activities that put him at risk"), sleep or rest, exercising or "activity to stay in shape" and diet as helpful management strategies and more discussed medication use. More of their children discussed many of these same coping strategies indicating that these are effective coping strategies that may lead to less intense perceptions of pain, less functional disability and better mental health. For example, one youth wrote "I've learned that I distract my mind from the pain by doing many activities just altering them to my own pace in which I can keep up." Many of these strategies have been shown to improve psychosocial functioning. For instance, by setting limits and promoting activity pacing, a child may feel less anxious about being able to meet expectations that were previously unattainable. Adequate sleep (Kahn et al., 2013), engaging in physical activity (Hearing et al., 2016) and eating a healthy diet (O'Neil et al., 2014)

are linked to positive mental health outcomes including reduced anxiety and depression. Parents without pain history and their children may not have identified these helpful strategies yet.

The patterns of qualitative responses between parents with and without pain and their children may also be reflective of what time phase of illness families are in. Parents without pain and their children, often discussed that having a diagnosis and knowledge, specifically, “knowing that there's a name to her pain and a cause to work with” makes living with hEDS easier. This mindset is reflective of an earlier stage of adaptation to chronic illness (i.e., crisis period) (Rolland, 1987). In contrast, parents and children in the positive parent pain history group rarely discussed these themes. Instead parent and child responses were focused on finding ways to cope with and maintain functioning within the child’s normal daily life, which is reflective of a later illness phase (i.e., chronic “long haul”) (Rolland, 1987). Families with parents with their own chronic pain history likely move into the chronic phase of illness sooner and are on a quicker path towards acceptance and management of their child’s condition due to the parents own experience adjusting to the “day-to-day living with a chronic illness” (Rolland, 1987). Thus having a diagnosis and gaining knowledge of the condition may not be as relevant for these families.

In addition to examining psychosocial and physical functioning differences, demographic differences between parent pain history groups were examined. Significant age differences were found. There was a greater range in age of children in the positive parent pain history group (i.e., 8-18 years old) and the inclusion of younger children, relative to the no parent pain history group (i.e., 13-17 years old). On average, children in the positive parent pain history group were also diagnosed two years younger (i.e. 10.19 years old vs 12.33 years old). Earlier diagnosis and receiving care at a pediatric genetic clinic at an earlier age may reflect that parents with a

positive pain history take their children to providers at an earlier age due to symptom identification or severity, leading to initiation of the diagnostic process sooner. Older age at diagnosis for children of parents without a pain history aligns with the typical experience of delay in diagnosis of hEDS (Castori et al., 2010; Kole & Faurisson, 2009). Many families see several providers over the course of years with misdiagnoses before they receive a diagnosis of hEDS (Castori et al., 2010). Lack of personal experience, may put families at a disadvantage for finding appropriate care sooner and contribute to delayed diagnosis as parents may not know which providers to take their child to until later in the diagnostic process.

Strengths and Limitations

The current study has many strengths including the inclusion of both parent and child report which allowed for the retention of individual perspectives of personal experience and the examination of relationships between parent and child variables. The inclusion of qualitative reports enriched the quantitative findings and provided potential explanations for findings. Finally, by collecting parent pain history, we were able to look at psychosocial transgenerational transmission factors which has not previously been examined within hEDS research.

In terms of study limitations, sample representativeness was a concern for generalizability of findings. Our sample was relatively small which limited the types of analyses we ran and what conclusions we could draw from findings. Small sample size contributed to non-significant t-test findings; however, we looked at effect sizes in addition to statistical significance to account for the impact of our small sample size on tests of statistical significance. While our sample was representative in terms of race and ethnicity, sex and income relative to other pediatric chronic pain research, it is not representative of the greater population. Our sample was a majority non-Hispanic White and so findings may not represent the experiences of

families of color. Sex differences within the positive parent pain history group highlight that boys and girls may experience their chronic pain and are impacted by their parents' pain experience differently. Participants were recruited solely from a pediatric genetics clinic, so our findings may not be generalizable as this setting may draw only a subsample of affected families. For example, all sample families had private insurance, which may indicate that families with public or no insurance are not able to access a genetics clinic. Relatedly, our sample contained a majority higher income families, which may represent a protective factor that contributes to more positive outcomes for families with a positive parent pain history. For families with higher socioeconomic status (SES) family health status may be the primary family stressor and thus, the family can devote more resources to supporting the health of family members. However, lower SES families have fewer resources and access to comprehensive care and numerous additional family stressors to contend with. Therefore, the potential positive impact of having a parent with chronic pain experience may not be as readily realized in families with lower SES or those without private insurance.

Additional limits center on the data collected. Data was collected from only one parent. Having both parents or caregivers would allow for greater examination of parental impact on child pain experience. For instance, within families in which the participating parent reported no pain history, other family members in the home may have hEDS or chronic pain and therefore, there may still be an influence by someone with a positive pain history impacting the child with hEDS. Additionally, as described earlier, we collected data on a limited number of parent factors. In this sample we assumed parents with chronic pain or hEDS were experiencing challenges that negatively impact their parenting, based on past research (De Baets et al., 2017; Wilson & Fales, 2015), but we did not collect measures of these potential challenges. Of note, the current

researchers attempted to improve upon many of these weaknesses by expanding study measures to include additional parent factors including parent functioning and recruit a larger more diverse sample by recruiting online through multiple social media platforms. Unfortunately, this method was unsuccessful, reflecting a broader challenge in conducting research with this population. In person recruitment of large samples is also a challenge as there is no one specialty that cares for patients with hEDS exclusively.

Clinical Implications

Previous literature prepared us to look at parent pain history as a significant risk factor for children's pain outcomes and while there is strong evidence of this, our findings suggest that this history may also offer some benefits to the management of children's pain. Therefore, the field may benefit from taking a strengths-based approach to family factors, rather than solely a risk factor based approach. Just as parents' own experience with chronic pain or hEDS is a potential resource for their child with hEDS, parents with personal experience would also be resources to other families. Consequently, providers should encourage and arrange for families of children with hEDS to join multifamily therapy, parent groups or child focused groups that include families with diverse chronic pain experiences or create a buddy or mentor program to pair less experienced families with a family who has multigenerational experience with hEDS. One parent from the no pain history group alluded to the benefit of other's experiences. They wrote that "reading inspiring blogs" makes living with EDS easier as "they give suggestions of things that work for some people." Equipping families with knowledge of the course of the illness as early as possible can help families prepare for and cope with future changes in functioning. However, because of the potential limits to linking families to each other for support (e.g., exacerbating each other's fears and anxiety or the sharing of non-medical expert advice) psychoeducation and

training for mentors and group facilitators should be integrated. Interventions for families across parent pain histories are also needed as physical and psychosocial functioning was negatively impacted in our overall sample. Access to multidisciplinary biopsychosocial rehabilitation and functional restoration programs to improve functioning is necessary for many youths (Friedrichsdorf et al., 2016). Components of this approach may include psychoeducation about hEDS and pain, physical therapy rehabilitation, integrative medicine/ active mind-body techniques, cognitive behavioral therapy, normalizing daily school attendance, sports, social life and sleep, parent coaching and medications (Friedrichsdorf et al., 2016). When thinking about what treatments would be most helpful and feasible for a family, providers need to consider broader family pain history and parent functioning as intervention for parent physical or mental health challenges may also be needed in order to maximize a child's ability to implement treatment recommendations (Cordts et al., 2019).

References

- Beighton, P., Paepe, A. D., Steinmann, B., Tsipouras, P., & Wenstrup, R. J. (1998). Ehlers-Danlos syndromes: revised nosology, Villefranche, 1997. *American journal of medical genetics*, 77(1), 31-37.
- Braun, V., & Clarke, V. (2006). Using thematic analysis in psychology. *Qualitative research in psychology*, 3(2), 77-101.
- Castarlenas, E., Jensen, M. P., von Baeyer, C. L., & Miró, J. (2017). Psychometric properties of the numerical rating scale to assess self-reported pain intensity in children and adolescents. *The Clinical journal of pain*, 33(4), 376-383.
- Castori, M., Camerota, F., Celletti, C., Danese, C., Santilli, V., Saraceni, V. M., & Grammatico, P. (2010). Natural history and manifestations of the hypermobility type Ehlers–Danlos syndrome: A pilot study on 21 patients. *American Journal of Medical Genetics Part A*, 152(3), 556-564.
- Castori, M., Dordoni, C., Valiante, M., Sperduti, I., Ritelli, M., Morlino, S., . . . Camerota, F. (2014). Nosology and inheritance pattern (s) of joint hypermobility syndrome and Ehlers-Danlos syndrome, hypermobility type: A study of intrafamilial and interfamilial variability in 23 Italian pedigrees. *American Journal of Medical Genetics Part A*, 164(12), 3010-3020.
- Claar, R. L., Simons, L. E., & Logan, D. E. (2008). Parental response to children's pain: The moderating impact of children's emotional distress on symptoms and disability. *Pain*, 138(1), 172-179. doi:10.1016/j.pain.2007.12.005
- Claar, R. L., & Walker, L. S. (2006). Functional assessment of pediatric pain patients: psychometric properties of the functional disability inventory. *Pain*, 121(1), 77-84.

- Cordts, K. M. P., Stone, A. L., Beveridge, J. K., Wilson, A. C., & Noel, M. (2019). The (parental) whole is greater than the sum of its parts: A multifactorial model of parent factors in pediatric chronic pain. *The Journal of Pain*.
- Creswell J and Poth C. (2017) *Qualitative Inquiry And Research Design Choosing Among Five Approaches* SAGE Publications, Inc.
- Crombez, G., Bijttebier, P., Eccleston, C., Mascagni, T., Mertens, G., Goubert, L., & Verstraeten, K. (2003). The child version of the pain catastrophizing scale (PCS-C): A preliminary validation. *Pain*, 104(3), 639-646.
- Cunningham, N. R., Lynch-Jordan, A., Barnett, K., Peugh, J., Sil, S., Goldschneider, K., & Kashikar-Zuck, S. (2014). Child pain catastrophizing mediates the relationship between parent responses to pain and disability in youth with functional abdominal pain. *Journal of pediatric gastroenterology and nutrition*, 59(6), 732.
- De Baets, S., Vanhalst, M., Coussens, M., Rombaut, L., Malfait, F., Van Hove, G., . . . Van de Velde, D. (2017). The influence of Ehlers-Danlos syndrome–hypermobility type, on motherhood: A phenomenological, hermeneutical study. *Research in developmental disabilities*, 60, 135-144.
- Denk, F., McMahon, S. B., & Tracey, I. (2014). Pain vulnerability: a neurobiological perspective. *Nature neuroscience*, 17(2), 192.
- Engelbert, R. H., Juul-Kristensen, B., Pacey, V., De Wandele, I., Smeenk, S., Woinarosky, N., . . . Simmonds, J. V. (2017). *The evidence-based rationale for physical therapy treatment of children, adolescents, and adults diagnosed with joint hypermobility syndrome/hypermobility Ehlers Danlos syndrome*. Paper presented at the American Journal of Medical Genetics Part C: Seminars in Medical Genetics.

- Fatoye, F., Palmer, S., Macmillan, F., Rowe, P., & van der Linden, M. (2012). Pain intensity and quality of life perception in children with hypermobility syndrome. *Rheumatology international*, 32(5), 1277-1284.
- Friedrichsdorf, S., Giordano, J., Desai Dakoji, K., Warmuth, A., Daughtry, C., & Schulz, C. (2016). Chronic pain in children and adolescents: diagnosis and treatment of primary pain disorders in head, abdomen, muscles and joints. *Children*, 3(4), 42.
- Goubert, L., Eccleston, C., Vervoort, T., Jordan, A., & Crombez, G. (2006). Parental catastrophizing about their child's pain. The parent version of the Pain Catastrophizing Scale (PCS-P): a preliminary validation. *Pain*, 123(3), 254-263.
- Hearing, C., Chang, W., Szuhany, K., Deckersbach, T., Nierenberg, A., & Sylvia, L. G. (2016). Physical exercise for treatment of mood disorders: a critical review. *Current behavioral neuroscience reports*, 3(4), 350-359.
- Irwin, D. E., Stucky, B., Langer, M. M., Thissen, D., DeWitt, E. M., Lai, J.-S., . . . DeWalt, D. A. (2010). An item response analysis of the pediatric PROMIS anxiety and depressive symptoms scales. *Quality of Life Research*, 19(4), 595-607.
- Kahn, M., Sheppes, G., & Sadeh, A. (2013). Sleep and emotions: bidirectional links and underlying mechanisms. *International Journal of Psychophysiology*, 89(2), 218-228.
- Kashikar-Zuck, S., Flowers, S. R., Claar, R. L., Guite, J. W., Logan, D. E., Lynch-Jordan, A. M., . . . Wilson, A. C. (2011). Clinical utility and validity of the Functional Disability Inventory (FDI) among a multicenter sample of youth with chronic pain. *Pain*, 152(7), 1600-1607.
- Kole, A., & Faurisson, F. (2009). The Voice of 12,000 Patients-Experiences and Expectations of Rare Disease Patients on Diagnosis and Care in Europe.

- Langer, S. L., Romano, J. M., Levy, R. L., Walker, L. S., & Whitehead, W. E. (2009). Catastrophizing and Parental Response to Child Symptom Complaints. *Children's Health Care, 38*(3), 169-184. doi:10.1080/02739610903038750
- Levy, R. L. (2010). Exploring the intergenerational transmission of illness behavior: from observations to experimental intervention. *Annals of Behavioral Medicine, 41*(2), 174-182.
- Lynch-Jordan, A. M., Kashikar-Zuck, S., Szabova, A., & Goldschneider, K. R. (2013). The interplay of parent and adolescent catastrophizing and its impact on adolescents' pain, functioning, and pain behavior. *The Clinical journal of pain, 29*(8), 681.
- Mu, W., Muriello, M., Clemens, J. L., Wang, Y., Smith, C. H., Tran, P. T., . . . Bodurtha, J. (2019). Factors affecting quality of life in children and adolescents with hypermobile Ehlers-Danlos syndrome/hypermobility spectrum disorders. *American Journal of Medical Genetics Part A, 179*(4), 561-569.
- Mulvey, M. R., Macfarlane, G. J., Beasley, M., Symmons, D. P., Lovell, K., Keeley, P., . . . McBeth, J. (2013). Modest Association of Joint Hypermobility With Disabling and Limiting Musculoskeletal Pain: Results From a Large-Scale General Population-Based Survey. *Arthritis care & research, 65*(8), 1325-1333.
- Noel, M., Palermo, T. M., Essner, B., Zhou, C., Levy, R. L., Langer, S. L., . . . Walker, L. S. (2015). A developmental analysis of the factorial validity of the parent-report version of the Adult Responses to Children's Symptoms in children versus adolescents with chronic pain or pain-related chronic illness. *The Journal of Pain, 16*(1), 31-41.

- O'Neil, A., Quirk, S. E., Housden, S., Brennan, S. L., Williams, L. J., Pasco, J. A., . . . Jacka, F. N. (2014). Relationship between diet and mental health in children and adolescents: a systematic review. *American journal of public health, 104*(10), e31-e42.
- Pacey, V., Tofts, L., Adams, R. D., Munns, C. F., & Nicholson, L. L. (2015). Quality of life prediction in children with joint hypermobility syndrome. *Journal of paediatrics and child health, 51*(7), 689-695.
- Palermo, T. M., & Chambers, C. T. (2005). Parent and family factors in pediatric chronic pain and disability: an integrative approach. *Pain, 119*(1-3), 1-4.
doi:10.1016/j.pain.2005.10.027
- Pielech, M., Ryan, M., Logan, D., Kaczynski, K., White, M. T., & Simons, L. E. (2014). Pain catastrophizing in children with chronic pain and their parents: Proposed clinical reference points and reexamination of the Pain Catastrophizing Scale measure. *PAIN®, 155*(11), 2360-2367.
- Rolland, J. S. (1987). Chronic illness and the life cycle: A conceptual framework. *Family process, 26*(2), 203-221.
- Scheper, M. C., Juul-Kristensen, B., Rombaut, L., Rameckers, E. A., Verbunt, J., & Engelbert, R. H. (2016). Disability in adolescents and adults diagnosed with hypermobility-related disorders: a meta-analysis. *Archives of physical medicine and rehabilitation, 97*(12), 2174-2187.
- Stone, A. L., & Wilson, A. C. (2016). Transmission of risk from parents with chronic pain to offspring: an integrative conceptual model. *Pain, 157*(12), 2628.
- Tinkle, B., Castori, M., Berglund, B., Cohen, H., Grahame, R., Kazkaz, H., & Levy, H. (2017). *Hypermobility Ehlers–Danlos syndrome (aka Ehlers–Danlos syndrome Type III and*

Ehlers–Danlos syndrome hypermobility type): *Clinical description and natural history*.

Paper presented at the American Journal of Medical Genetics Part C: Seminars in Medical Genetics.

Trost, Z., Strachan, E., Sullivan, M., Vervoort, T., Avery, A. R., & Afari, N. (2015). Heritability of pain catastrophizing and associations with experimental pain outcomes: a twin study. *Pain, 156*(3), 514.

Van Slyke, D. A., & Walker, L. S. (2006). Mothers' responses to children's pain. *The Clinical journal of pain, 22*(4), 387-391.

Varni, J. W., Magnus, B., Stucky, B. D., Liu, Y., Quinn, H., Thissen, D., . . . DeWalt, D. A. (2014). Psychometric properties of the PROMIS® pediatric scales: precision, stability, and comparison of different scoring and administration options. *Quality of Life Research, 23*(4), 1233-1243.

Walker, L. S., Garber, J., & Greene, J. W. (1991). Somatization symptoms in pediatric abdominal pain patients: relation to chronicity of abdominal pain and parent somatization. *Journal of abnormal child psychology, 19*(4), 379-394.

Walker, L. S., & Zeman, J. L. (1992). Parental response to child illness behavior. *Journal of pediatric psychology, 17*(1), 49-71.

Welkom, J. S., Hwang, W.-T., & Guite, J. W. (2013). Adolescent pain catastrophizing mediates the relationship between protective parental responses to pain and disability over time. *Journal of pediatric psychology, jst011*.

Wilson, A. C., & Fales, J. L. (2015). Parenting in the context of chronic pain: a controlled study of parents with chronic pain. *The Clinical journal of pain, 31*(8), 689.

Wilson, A. C., Moss, A., Palermo, T. M., & Fales, J. L. (2014). Parent pain and catastrophizing are associated with pain, somatic symptoms, and pain-related disability among early adolescents. *Journal of Pediatric Psychology, 39*(4), 418-426.

Table 1.

Demographic Characteristics of Participants

	Children and Adolescents						Caregivers					
	Whole Sample		Positive Parent Pain History		No Parent Pain History		Whole Sample		Positive Parent Pain History		No Parent Pain History	
	Mean (SD)	Range	Mean (SD)	Range	Mean (SD)	Range	Mean (SD)	Range	Mean (SD)	Range	Mean (SD)	Range
Age	14.7 (2.9)	8-18	13.8 (3.4)	8-18	15.8 (1.4)	13-17	46.5 (7.6)	32-66	46.1 (7.4)	32-56	47.1 (8.1)	34-66
	<i>n</i>	%	<i>n</i>	%	<i>n</i>	%	<i>n</i>	%	<i>n</i>	%	<i>n</i>	%
Sex												
Female	24	70.6	9	56.3	11	91.7	26	92.9	15	93.8	11	91.7
Male	8	23.5	6	37.5	1	8.3	2	7.1	1	6.3	1	8.3
Not Reported	2	5.9	1	6.3	0	0	0	0	0	0	0	0
Race/Ethnicity												
non-Hispanic White	18	52.9	11	68.8	7	58.3	23	82.1	14	87.5	9	75
Hispanic	7	20.6	5	31.2	2	16.7	2	7.1	2	12.5	0	0
Asian or Asian American	0	0	0	0	0	0	2	7.1	0	0	2	16.7
Biracial	3	8.8	0	0	3	25	1	3.6	0	0	1	8.3
Asian or Asian American and White	2	5.9	0	0	2	16.7	0	0	0	0	0	0
American Indian or Alaska Native and White	1	2.9	0	0	1	8.3	1	3.6	0	0	1	8.3
Not Reported	6	17.6	0	0	0	0	0	0	0	0	0	0
Relationship to Child												
Mother	-	-	-	-	-	-	25	89.3	15	93.8	10	83.3
Father							2	7.1	1	6.2	1	8.3
Grandmother							1	3.6	0	0	1	8.3
Family Income												
\$25,000 - \$50,000							2	7.1	2	12.5	0	0
\$50,000 - \$75,000							3	10.7	1	6.3	2	16.7
\$75,000 - \$100,000							7	25.0	5	31.3	2	16.7
\$100,000 - \$150,000							8	28.6	3	18.8	5	41.7
More than \$150,000							6	21.4	4	25.0	2	16.7
Not Reported							2	7.1	1	6.3	1	8.3

Table 2.

Correlational Table of Study Variables

Variable	n	1	2	3	4	5	6
1. Pain Level	34	-					
2. Functional Disability	34	.64***	-				
3. Anxiety	34	.47***	.56***	-			
4. Depression	34	.48***	.57***	.79***	-		
5. Child Pain Catastrophizing	33	.45***	.49***	.58***	.69***	-	
6. Parent Pain Catastrophizing	27	.20	.23	.35*	.11	.16	-
7. Protectiveness	28	-.03	-.09	-.22	-.31	-.07	.00

Note. Variables 1 through 5 are child variables and 6 and 7 are parent variables. * $p < .1$. ** $p < .05$. *** $p < .01$.

Table 3.

Main Analyses

Variable	Parent Pain History	Descriptives			Independent t-tests			Effect Size
		N	Mean	SD	t	df	p	Hedge's G
Pain Level	No Pain	12	6.00	2.26	1.50	25.30	0.146	0.55
	Positive Pain	16	4.63	2.58				
Functional Disability	No Pain	12	21.68	6.36	1.15	22.25	0.261	0.39
	Positive Pain	16	17.16	13.83				
Anxiety	No Pain	12	57.32	12.19	1.56	21.99	0.134	0.59
	Positive Pain	16	50.44	10.69				
Depression	No Pain	12	55.56	11.41	1.54	24.81	0.137	0.56
	Positive Pain	16	48.61	12.37				
Child Pain Catastrophizing	No Pain	12	23.00	9.49	1.46	24.94	0.157	0.54
	Positive Pain	15	17.13	11.39				
Parent Pain Catastrophizing	No Pain	12	21.75	11.35	0.917	22.67	0.369	0.35
	Positive Pain	15	17.87	10.38				
Protectiveness	No Pain	12	19.65	3.56	0.740	20.30	0.468	0.25
	Positive Pain	16	17.76	9.37				

Note. Variables 1 through 5 are child variables and 6 and 7 are parent variables. * $p < .1$. ** $p < .05$. *** $p < .01$.

Table 4.

Qualitative Responses to “What Makes Having EDS Easier?”

Codes: What Makes Having EDS Easier?	Children and Adolescents		Parents	
	No Parent Pain History N = 9	Positive Parent Pain History N = 13	No Pain History N = 10	Positive Pain History N = 12
Positive physical attributes of having hEDS (e.g., flexible, tall & thin)	1 (11.1)	1 (7.7)	0 (0)	0 (0)
Own understanding/knowledge of hEDS/ Having a diagnosis	2 (22.2)	0 (0)	4 (40)	1 (8.3)
Others’ understanding of hEDS	1 (11.1)	1 (7.7)	0 (0)	0 (0)
Social Support	1 (11.1)	2 (15.4)	3 (30)	2 (16.7)
Quality medical care/ Support from medical professionals	0 (0)	0 (0)	4 (40)	5 (41.7)
School accommodations/ School support	0 (0)	0 (0)	3 (30)	0 (0)
Activity Pacing/ Setting Limits	0 (0)	2 (15.4)	0 (0)	2 (16.7)
Sleep/Rest	2 (22.2)	2 (15.4)	0 (0)	4 (33.3)
Distraction/Keeping busy	1 (11.1)	1 (7.7)	0 (0)	0 (0)
Exercise(s)/ Staying active	1 (11.1)	4 (30.8)	0 (0)	3 (25)
Physical Therapy/ Orthotics/Braces	3 (33.3)	2 (15.4)	3 (30)	3 (25)
Medication	2 (22.2)	4 (30.8)	1 (10)	3 (25)
Dietary management	1 (11.1)	0 (0)	0 (0)	2 (16.7)
Complementary/ Integrative techniques (e.g., heat/ice, physical manipulation, Cannabidiol, massage, Epsom salt)	2 (22.2)	3 (23.1)	3 (30)	3 (25)

Note. Number of individual people who mentioned the given code (percent of members of that group (e.g. children and adolescents in the no parent pain history group) who mentioned given code)

Appendix A. Measures

Parental Pain History

Parent Pain History group was determined by caregiver response to the questions:

“Other family member with EDS”

“Other family members with chronic pain (of any type) and what type”

If the parent indicated that they had EDS or a chronic pain condition, their family was allocated into the positive parent pain history group. If they did not indicate that they had EDS or another chronic pain condition, their family was allocated into the no parent pain history group.

Pain Catastrophizing Scale Parent Version (PCS-P)

PCS-P

Thoughts and feelings when you child is in pain

We are interested in the thoughts and feelings you have when your child is in pain. Below are 13 sentences of different thoughts and feelings. Please put a circle around the word or phrase under each sentence that best reflects how strongly you have each thought when your child is in pain.

1. When my child is in pain, I worry all the time about whether the pain will end.
NOT AT ALL MILDLY MODERATELY SEVERELY EXTREMELY
2. When my child is in pain, I feel I can't go on like this much longer.
NOT AT ALL MILDLY MODERATELY SEVERELY EXTREMELY
3. When my child is in pain, it's terrible and I think it's never going to get better.
NOT AT ALL MILDLY MODERATELY SEVERELY EXTREMELY
4. When my child is in pain, it's awful and I feel that it overwhelms me
NOT AT ALL MILDLY MODERATELY SEVERELY EXTREMELY
5. When my child is in pain, I can't stand it anymore
NOT AT ALL MILDLY MODERATELY SEVERELY EXTREMELY
6. When my child is in pain, I become afraid that the pain will get worse
NOT AT ALL MILDLY MODERATELY SEVERELY EXTREMELY
7. When my child is in pain, I keep thinking of other painful events
NOT AT ALL MILDLY MODERATELY SEVERELY EXTREMELY
8. When my child is in pain, I want the pain to go away
NOT AT ALL MILDLY MODERATELY SEVERELY EXTREMELY
9. When my child is in pain, I can't keep it out of my mind
NOT AT ALL MILDLY MODERATELY SEVERELY EXTREMELY
10. When my child is in pain, I keep thinking about how much he/she is suffering
NOT AT ALL MILDLY MODERATELY SEVERELY EXTREMELY
11. When my child is in pain, I keep thinking about how much I want the pain to stop
NOT AT ALL MILDLY MODERATELY SEVERELY EXTREMELY
12. When my child is in pain, there is nothing I can do to stop the pain.
NOT AT ALL MILDLY MODERATELY SEVERELY EXTREMELY
13. When my child is in pain, I wonder whether something serious may happen
NOT AT ALL MILDLY MODERATELY SEVERELY EXTREMELY

Adult Responses to Children's Symptoms (ARCS) Protect Subscale

ADULT RESPONSES TO CHILDREN'S SYMPTOMS (ARCS) Parent Form

What happens when your child is in pain?

The next questions are about what you do when your child is in pain. For each question, choose one of the answers:

Never	means that you never do this.
Once in a while	means that you only do this once in a while.
Sometimes	means that you do this some of the time.
Often	means that you usually do this.
Always	means that you always do this.

When your child is in pain, how often do you . . .

	<u>Never</u>	<u>Once in a while</u>	<u>Some- times</u>	<u>Often</u>	<u>Always</u>
1. Ask your child what you can do to help?	0	1	2	3	4
2. Express irritation or frustration with your child?	0	1	2	3	4
3. Do your child's chores or pick up your child's things instead of making him/her do it?	0	1	2	3	4
4. Talk to your child about something else to take your child's mind off it?	0	1	2	3	4
5. Give your child some medicine?	0	1	2	3	4
6. Reassure your child that he/she is going to be OK?	0	1	2	3	4
7. Get your child something to eat or drink?	0	1	2	3	4
8. Bring your child special treats or little gifts?	0	1	2	3	4
9. Try not to pay attention to your child?	0	1	2	3	4
10. Ask your child questions about how he/she feels?	0	1	2	3	4
11. Let your child stay home from school?	0	1	2	3	4
12. Encourage your child to do something he or she enjoys (like watch TV or play a game)?	0	1	2	3	4
13. Tell your child that he/she doesn't have to finish all of his/her homework?	0	1	2	3	4

**ADULT RESPONSES TO CHILDREN'S SYMPTOMS (ARCS)
Parent Form**

When your child is in pain, how often do you . . .

	<u>Never</u>	<u>Once in a while</u>	<u>Some- times</u>	<u>Often</u>	<u>Always</u>
14. Tell your child there's nothing you can do about it?	0	1	2	3	4
15. Give your child special privileges?	0	1	2	3	4
16. Stay home from work or come home early (or stay home instead of going out or running errands)?	0	1	2	3	4
17. Tell others in the family not to bother your child or to be especially nice to your child?	0	1	2	3	4
18. Tell your child not to make such a fuss about it?	0	1	2	3	4
19. Pay more attention to your child than usual?	0	1	2	3	4
20. Let your child sleep in a special place (like in your room or on the couch)?	0	1	2	3	4
21. Tell your child that he/she needs to learn to be stronger?	0	1	2	3	4
22. Let your child sleep later than usual in the morning?	0	1	2	3	4
23. Keep your child inside the house?	0	1	2	3	4
24. Try to involve your child in some activity?	0	1	2	3	4
25. Spend more time than usual with your child?	0	1	2	3	4
26. Try to make your child as comfortable as possible?	0	1	2	3	4
27. Tell your child you still expect him/her to do his/her chores or pick up his/her things around the house?	0	1	2	3	4
28. Check on your child to see how he/she is doing?	0	1	2	3	4
29. Call the doctor or take your child to the doctor?	0	1	2	3	4

Protect Subscale includes questions: 3, 7, 8, 11, 13, 15, 16, 17, 19, 20, 22, 23, 25

Pain Catastrophizing Scale Child Version (PCS-C)

PCS-C

Thoughts and feelings during pain

We are interested in what you think and how strong the feelings are when you are in pain. Below are 13 sentences of different thoughts and feelings you can have when you are in pain. Try to show us as clearly as possible what you think and feel by putting a circle around the word under each sentence that best reflects how strongly you have each thought.

- | | | | | | | |
|-----|--|------------|--------|------------|----------|-----------|
| 1. | When I am in pain, I worry all the time about whether the pain will end. | NOT AT ALL | MILDLY | MODERATELY | SEVERELY | EXTREMELY |
| 2. | When I am in pain, I feel I can't go on like this much longer. | NOT AT ALL | MILDLY | MODERATELY | SEVERELY | EXTREMELY |
| 3. | When I am in pain, it's terrible and I think it's never going to get better. | NOT AT ALL | MILDLY | MODERATELY | SEVERELY | EXTREMELY |
| 4. | When I am in pain, it's awful and I feel that it takes over me | NOT AT ALL | MILDLY | MODERATELY | SEVERELY | EXTREMELY |
| 5. | When I am in pain, I can't stand it anymore | NOT AT ALL | MILDLY | MODERATELY | SEVERELY | EXTREMELY |
| 6. | When I am in pain, I become afraid that the pain will get worse | NOT AT ALL | MILDLY | MODERATELY | SEVERELY | EXTREMELY |
| 7. | When I am in pain, I keep thinking of other painful events | NOT AT ALL | MILDLY | MODERATELY | SEVERELY | EXTREMELY |
| 8. | When I am in pain, I want the pain to go away | NOT AT ALL | MILDLY | MODERATELY | SEVERELY | EXTREMELY |
| 9. | When I am in pain, I can't keep it out of my mind | NOT AT ALL | MILDLY | MODERATELY | SEVERELY | EXTREMELY |
| 10. | When I am in pain, I keep thinking about how much it hurts | NOT AT ALL | MILDLY | MODERATELY | SEVERELY | EXTREMELY |
| 11. | When I am in pain, I keep thinking about how much I want the pain to stop | NOT AT ALL | MILDLY | MODERATELY | SEVERELY | EXTREMELY |
| 12. | When I am in pain, there is nothing I can do to stop the pain. | NOT AT ALL | MILDLY | MODERATELY | SEVERELY | EXTREMELY |
| 13. | When I am in pain, I wonder whether something serious may happen | NOT AT ALL | MILDLY | MODERATELY | SEVERELY | EXTREMELY |

Pain Intensity Scale

On days that you have had pain, what has been your usual level of pain in the last 2 weeks?

0	1	2	3	4	5	6	7	8	9	10
No Pain at All										Worst Pain I Can Imagine

Patient Reported Outcomes Measurement Information System (PROMIS) Pediatric

Anxiety Subscale

Please respond to each item by marking one box per row.

In the past 7 days...

	Never	Almost Never	Sometimes	Often	Almost Always
I felt like something awful might happen.					
I felt nervous.					
I felt scared.					
I felt worried.					
I worried when I was at home.					
I got scared really easy.					
I worried about what could happen to me.					
I worried when I went to bed at night.					

PROMIS Pediatric Depression Subscale

Please respond to each item by marking one box per row.

In the past 7 days...

	Never	Almost Never	Sometimes	Often	Almost Always
I could not stop feeling sad.					
I felt alone.					
I felt everything in my life went wrong.					
I felt like I couldn't do anything right.					
I felt lonely.					
I felt sad.					
I felt unhappy.					
It was hard for me to have fun.					

Functional Disability Inventory (FDI)

Functional Disability Inventory Child and Adolescent Form

When people are sick or not feeling well it is sometimes difficult for them to do their regular activities. In the past two weeks, would you have had any physical trouble or difficulty doing these activities?

	No Trouble	A Little Trouble	Some Trouble	A Lot of Trouble	Impossible
1. Walking to the bathroom.	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
2. Walking up stairs.	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
3. Doing something with a friend. (For example, playing a game.)	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
4. Doing chores at home.	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
5. Eating regular meals.	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
6. Being up all day without a nap or rest.	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
7. Riding the school bus or traveling in the car.	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4

Remember, you are being asked about difficulty due to physical health.

	No Trouble	A Little Trouble	Some Trouble	A Lot of Trouble	Impossible
8. Being at school all day.	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
9. Doing the activities in gym class (or playing sports).	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
10. Reading or doing homework.	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
11. Watching TV.	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
12. Walking the length of a football field.	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
13. Running the length of a football field.	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
14. Going shopping.	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4
15. Getting to sleep at night and staying asleep.	<input type="checkbox"/> 0	<input type="checkbox"/> 1	<input type="checkbox"/> 2	<input type="checkbox"/> 3	<input type="checkbox"/> 4

Codes Developed for Responses to “What makes living with EDS easier?”

Child Codes		Description and Example Responses
1	Positive physical attributes of having hEDS	Perceived positive aspects of physical appearance (being tall and thin) or ability (flexibility) as a result of having hEDS; “I get to use my flexibility in dance”
2	My own understanding of hEDS	Having a better understanding of hEDS and symptoms via education and awareness; “knowing that there is a reason why I keep getting injured”
3	Others understanding of hEDS	Others being knowledgeable about hEDS and demonstrating understanding; “finding others who relate/understand”
4	Medication	Medication; “pain medications”
5	Sleep/rest	Sleep and rest; “getting enough sleep”
6	Physical therapy (PT)	Participating in physical therapy; “strain counterstrain physical therapy”
7	Exercise/exercises/staying active	Exercising, doing exercises and staying active; “exercise”
8	Social support	Having support from friends and family; “the support of my friends/family”
9	Distraction/keeping busy	Keeping one’s mind off pain through distractions including a busy schedule; “...I distract my mind from the pain by doing many activities...”
10	Dietary management (homeostasis)	Engaging in dietary management and helping to maintain internal homeostasis like consuming enough sodium and liquids; “drinking enough fluids”
11	Complementary/integrative techniques/treatments	Engaging in complementary and integrative medicine techniques and treatments such as using a heating pad, icing, Cannabidiol (CBD), and physical manipulation other than physical therapy, like massage; “massages help”
12	Activity pacing/setting limits	Limiting or adapting activities to be more manageable; “having my limits respected”
13	I don’t know	Not knowing what is helpful or not providing a helpful strategy; “nothing I do makes me feel better”

Parent Codes		Example Responses
1	School accommodations/limiting activities/pacing	Limiting or adapting activities to be more manageable, including accommodations at school; “limited activities that put him at risk”
2	Sleep/rest	Sleep and rest; “sleep, rest...”
3	Medication	Medication; “medication”
4	Physical Therapy/orthotics/braces	Participating in physical therapy and utilizing other physical mobility supports like orthotics and braces; “he has done PT”

5	Having a diagnosis/ knowledge about the condition	Having a diagnosis and knowledge of the condition and symptoms; “I believe that knowledge of the symptoms and syndrome”
6	Exercise/staying active	Exercising and staying active; “activity to stay in shape”
7	Diet	Engaging in dietary management and helping to maintain internal homeostasis like consuming enough liquids; “maintaining proper diet”
8	Social support	Having support from friends and family; “support of loved ones”
9	Quality medical care/support from medical professionals	Perceiving good quality medical care (e.g., knowledgeable doctors) and supportive and understanding medical professionals; “finding the DR's that can help, not telling you it in your head”
10	Complementary/integrative techniques/treatments	Engaging in complementary and integrative medicine techniques and treatments such as using a heating pad, icing, Cannabidiol (CBD), and physical manipulation other than physical therapy, like massage and; “medical marijuana, CBD”

Appendix B. Proposal

Influence of Parent Chronic Pain on
Youth's Experience of Hypermobile Ehlers-Danlos Syndrome

Proposal for a Thesis

Presented to

The Department of Psychology

DePaul University

By

Marissa Lee Koven

October 25, 2018

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Abstract

This thesis examines parent factors that may relate to youth's experiences with Hypermobile Ehlers Danlos Syndrome (hEDS). Youth with hEDS are at risk for physical limitations and psychological distress associated with pain and other symptomology. The greater pediatric chronic pain and transgenerational pain risk literature draws attention to multiple pathways through which parents influence their children's psychological and other pain-related outcomes; however, the relationship between parental hEDS experience and child hEDS experience has yet to be explored. Due to the substantial impact that chronic pain and hEDS may have on a parent's own psychological, physical functioning and parenting, it is possible that they think about pain and respond to their children's pain complaints differently than parents without a history of pain. This thesis presents a model of transgenerational transmission of risk associated with chronic pain. It is hypothesized that parents with chronic pain will be more likely to catastrophize about pain and respond to their child's pain more protectively than parents without their own history of pain. Parental catastrophic thinking about pain is hypothesized to be passed onto their child through the use of protective response behaviors, leading the child to catastrophize about pain. Parents who catastrophize about their child's pain more and use more protective response behaviors are hypothesized to have children with increased pain intensity, functional disability, and symptoms of anxiety and depression. In order to investigate the proposed relationships, parent-child pairs completed self-report measures that assessed a variety of pain related factors including chronic pain diagnoses, beliefs about pain, parental response behaviors and child pain-related outcomes. It is clinically important to identify risks for worse pain-related child outcomes and the pathway through which risk is transmitted, in order to design targeted family treatment and prevention programs that minimize risk for families affected by hEDS.

Introduction

The current study examines parent factors that may relate to youth's experiences with Hypermobile Ehlers Danlos Syndrome (hEDS). HEDS, its symptomology, and associated psychosocial and physical outcomes will be reviewed. A model of the transgenerational transmission of risk associated with chronic pain is presented. Parent's own experience with chronic pain is highlighted as an important determinant of how parents think about and respond to their child's pain. Potential pathways through which parent factors influence a child's own thinking about pain are investigated. The goal of the study is to learn more about parent factors that influence child pain-related outcomes and the pathways through which they exert their influence. A greater understanding of the transmission of risk within families affected by hEDS can inform future intervention and treatment for youth with hEDS to increase their efficacy and lead to more positive pain-related outcomes.

HEDS

The Ehlers Danlos Syndromes (EDS) are a set of heritable connective tissue disorders (Malfait et al., 2017). HEDS is the most common of the EDS subtypes and likely accounts for 80 to 90% of all EDS cases (Tinkle et al., 2017), affecting about 255 million people worldwide (Mulvey et al., 2013; Tinkle et al., 2017). Many hEDS symptoms negatively impact physical and psychosocial functioning which put individuals at risk for functional disability, anxiety, depression, and reduced health related quality of life (HRQoL) (Bulbena et al., 2017; Castori et al., 2012; Fatoye, Palmer, Macmillan, Rowe, & van der Linden, 2012; Pacey, Tofts, Adams, Munns, & Nicholson, 2015; Scheper et al., 2016; Sinibaldi, Ursini, & Castori, 2015; Tinkle et al., 2017).

In order for a diagnosis of hEDS to be made, an individual must meet three criteria. First, they must show signs of Generalized Joint Hypermobility (GJH), which involves a Beighton score of joint hyperflexibility of at least 5 out of 9 (Malfait et al., 2017). It is recommended that pre-pubertal children and adolescents exceed a Beighton score of 6, since children and adolescents are normatively more flexible than adults (Malfait et al., 2017). Second, an individual must have at least two of the following: systemic manifestations of a connective tissue disorder (e.g. velvety soft skin, hyperextendible (stretchy) skin, or unexplained stretch marks); a history of at least one first degree relative diagnosed with hEDS; and/or musculoskeletal complications that may include chronic (occurring for at least three months) daily musculoskeletal pain in two or more limbs, chronic widespread pain, recurrent joint dislocations or joint instability (Malfait et al., 2017). Third, a diagnosis of hEDS is made once other forms of EDS, other connective tissue disorders, and alternative diagnoses that explain the hypermobile joints have been ruled out (Malfait et al., 2017).

It has been proposed that hEDS can be described in terms of three “discrete” disease phases: hypermobility, pain, and stiffness (Tinkle et al., 2017). The Hypermobility Phase begins at an early age and is marked by extreme flexibility (Tinkle et al., 2017), which may help children excel at sports like gymnastics or dance, but children with hEDS also commonly experience an increased incidence of sprains, dislocations, fatigue, and pain (Tinkle et al., 2017). The Pain Phase typically begins somewhere between adolescence and middle-age and is characterized by chronic and progressively widespread musculoskeletal pain (Tinkle et al., 2017). There is also typically a worsening of fatigue, additional forms of chronic pain like headaches as well as more systemic effects (Tinkle et al., 2017). The Stiffness Phase is

characterized by generalized reduction of joint mobility, significant functional disability due to pain, fatigue, and other symptoms causing motor limitations (Tinkle et al., 2017).

Symptom presentation varies considerably even among children and adolescents who experience the hypermobility and pain phases. Across a sample of youth with Joint Hypermobility Syndrome (JHS), which is considered to be clinically indistinguishable from hEDS (Tinkle et al., 2009), Pacey and colleagues identified five distinct subtypes of JHS presentations: Joint Affected, Athletic, Systemic, Soft Tissue Affected, and High BMI (Pacey, Adams, Tofts, Munns, & Nicholson, 2014). More generally, they found that a large majority (91%) of children and adolescents in the study had non-musculoskeletal involvement including (in order from highest to lowest incidence) skin, gastrointestinal, cardiovascular, eye, incontinence, and hernia (Pacey et al., 2014). It was also more common for chronic pain to be present in accordance with recurrent joint instability (61%) than for chronic pain to be present alone (31%) or recurrent joint instability alone (8%) (Pacey et al., 2014).

HEDS Outcomes

Children and adolescents with hEDS are at risk for functional impairment (Adib, Davies, Grahame, Woo, & Murray, 2005), psychological distress (Engelbert et al., 2017) and reduced HRQoL (Cattalini, Khubchandani, & Cimaz, 2015; Fatoye et al., 2012; Pacey et al., 2014; Pacey et al., 2015). Functional impairment may include difficulty with writing tasks and various physical activities (i.e. physical education activities, sports, outdoor games, riding a bicycle) (Adib et al., 2005; Schubert-Hjalmarsson, Öhman, Kyllerman, & Beckung, 2012). Psychological distress has only been looked at broadly; it may include poorer emotional (Fatoye et al., 2012; Pacey et al., 2015) or psychosocial functioning and self-esteem (Pacey, Tofts, Adams, Munns, & Nicholson, 2013). HRQoL has been the most common psychosocial measure used with youth

with hEDS. For children with hEDS, every domain of HRQoL (i.e. physical, emotional, social and school) is at risk for being reduced, with physical functioning being the most substantially impacted domain (Fatoye et al., 2012; Pacey et al., 2015). Pacey and colleagues found that 75% of the variance in child-reported HRQoL can be accounted for by general fatigue, sleep and rest related fatigue, pain intensity and presence or absence of stress incontinence symptoms (Pacey et al., 2015). Reports of reduced HRQoL among children and adolescents with JHS indicate that these youths, like youths with other chronic pain and chronic illness like fibromyalgia, cancer, and obesity, are at risk for reduced HRQoL compared to healthy children (Pacey et al., 2015).

The literature on pediatric hEDS is limited compared to other chronic pain conditions, but it offers preliminary evidence that children with hEDS respond similarly to their pain experiences compared to youth with other chronic pain conditions (Fatoye et al., 2012; Pacey et al., 2015). Therefore, it is useful to look at psychosocial and physical outcomes in other pediatric chronic pain samples to learn more about how youth with hEDS may respond and adapt to their condition. Reduced HRQoL, functional disability, anxiety symptoms, depressive symptoms, social functioning and school disability and absenteeism have all been found to be prevalent concerns in various chronic pain populations including abdominal pain, headaches, musculoskeletal pain, juvenile idiopathic/rheumatoid arthritis, fibromyalgia, and chronic fatigue syndrome (Claar & Walker, 2006; Forgeron et al., 2010; Garralda & Rangel, 2004; Kashikar-Zuck et al., 2011; Kashikar-Zuck, Goldschneider, Powers, Vaught, & Hershey, 2001; Kashikar-Zuck et al., 2008; Logan, Simons, Stein, & Chastain, 2008; Pacey et al., 2015).

The adult hEDS literature provides additional support for the potential negative psychological impact of hEDS on children and adolescents. Almost half of adults with hEDS may suffer from a psychiatric disorder, with anxiety and depression being the most prevalent

(Bulbena et al., 2017; Hershenfeld et al., 2016). Anxiety has also been linked with higher levels of pain catastrophizing, somatosensory amplification (i.e. hypervigilance to somatic and internal systems related sensations), poorer social functioning, and perceived poor general health in adult hEDS patients (Baeza-Velasco et al., 2018). Additionally, adults with hEDS have been found to have lower quality of life related to physical pain, systemic effects like functional gastrointestinal disorders, fatigue, and psychological distress (Castori et al., 2012; Fikree, Chelimsky, Collins, Kovacic, & Aziz, 2017; Hakim, De Wandele, O'Callaghan, Pocinki, & Rowe, 2017; Krahe, Adams, & Nicholson, 2018; Malfait et al., 2017; Rombaut et al., 2011; Scheper et al., 2016; Tinkle et al., 2017).

Intergenerational Transmission of Risk Associated with Chronic Pain and hEDS

An important contributing factor to youth's hEDS experience is that hEDS is inherited, and so, children and adolescents with hEDS are likely to have a parent or other family members with hEDS (Castori et al., 2014). While various aspects of parent-child relationships have been explored in other pediatric chronic pain samples, only two hEDS studies have looked at parent-child variables (De Baets et al., 2017; Pacey et al., 2015) and none have examined the relationship between parent and child chronic pain experiences or the influence of parent-child interactions on child outcomes. Therefore, there is a need to fill this gap in the literature. The current study seeks to help fill this gap by examining patterns of disability and psychological functioning among pediatric hEDS patients in relation to parental chronic pain history, parent pain-related cognitions, and parent driven pain-specific social learning.

Stone and Wilson proposed a Conceptual Model of Intergenerational Transmission of Chronic Pain Risk (Stone & Wilson, 2016) (Figure 1) that I believe helps conceptualize parent-related sources of physical and psychosocial risks for children with hEDS. Stone and Wilson

discuss their model in terms of parental chronic pain as a risk factor for the development of pediatric chronic pain with multiple mechanisms for risk transmission. These mechanisms include genetics, early neurobiological development, pain-specific social learning, general parenting and family history, and exposure to a stressful environment (Stone & Wilson, 2016). These mechanisms are proposed to bidirectionally interact with child vulnerabilities which include pain processing, pain-related cognitions and affect, pain coping behaviors, physical health factors, and emotion regulation; which in turn influence pain-related child outcomes such as chronic pain experience, disability, and psychological functioning (Stone & Wilson, 2016). While Stone and Wilson focus on parents with a history of chronic pain, their proposed mechanisms would still interact with child vulnerabilities regardless of parental chronic pain history (Denk, McMahon, & Tracey, 2014; Palermo & Chambers, 2005). However, past research leads us to believe that the interaction between these mechanisms and child vulnerabilities may differ for children of parents with and without chronic pain (Palermo & Chambers, 2005; Wilson & Fales, 2015; Wilson, Moss, Palermo, & Fales, 2014) as parent's own chronic pain experiences, or lack thereof, inform mechanism pathways.

I have adapted Stone and Wilson's model to account for various parental chronic pain experiences by changing the risk from Parent with Chronic Pain to Parent Chronic Pain History which may include hEDS related pain, non-hEDS chronic pain, or no history of chronic pain. I further propose the insertion of Parent Vulnerabilities between Risk and Mechanisms to conceptualize parent variables influenced by their own chronic pain, or lack of chronic pain, experiences, that subsequently inform the Mechanisms that interplay with Child Vulnerabilities.

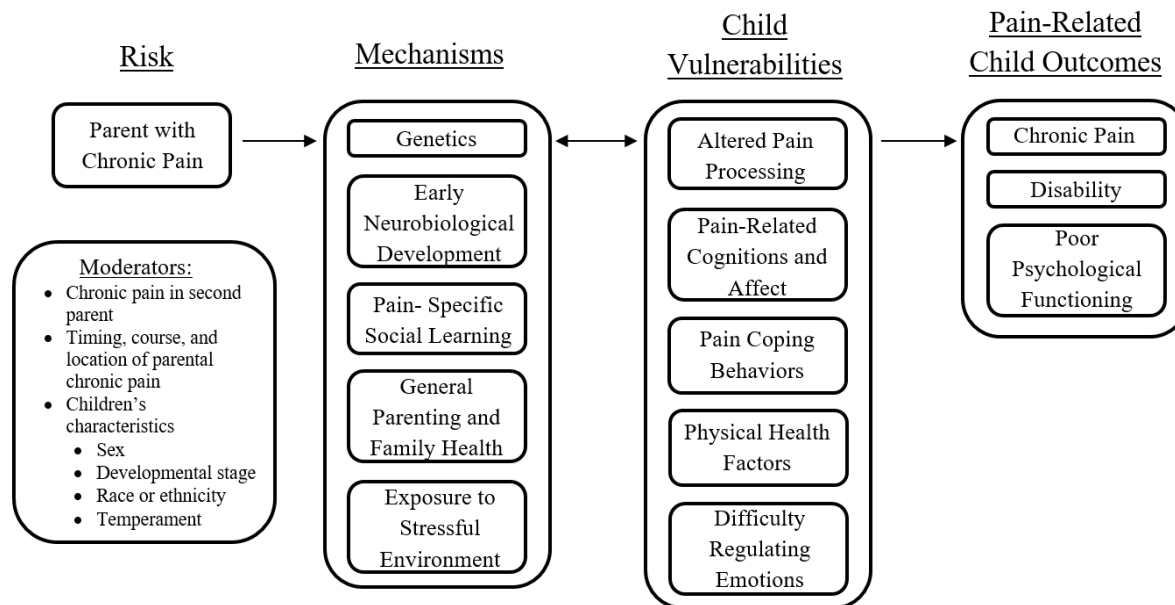


Figure 1. Conceptual Model of Intergenerational Transmission of Chronic Pain Risk (Stone & Wilson, 2016).

While there is a great breadth of factors that interact to impact Pain-Related Child Outcomes, the current study focused on a subset of variables and their relationships. Figure 2 demonstrates the adapted model and highlights which variables were measured in the current study. Parent history of chronic pain is likely to affect parent vulnerabilities such as how that parent thinks about their child's pain. This in turn is likely to influence the way that parent responds to their child's pain behaviors (mechanisms). A child's own beliefs about pain are likely influenced by parent beliefs, which they may learn through their parents' responses to their pain. How a child thinks about pain (child vulnerabilities) will impact their perception of pain, subsequent functional disability, and feelings of anxiety and depression (outcomes). The following sections will discuss research with hEDS and other pediatric chronic pain samples that pertains to the proposed relationships and pathway presented in the adapted model.

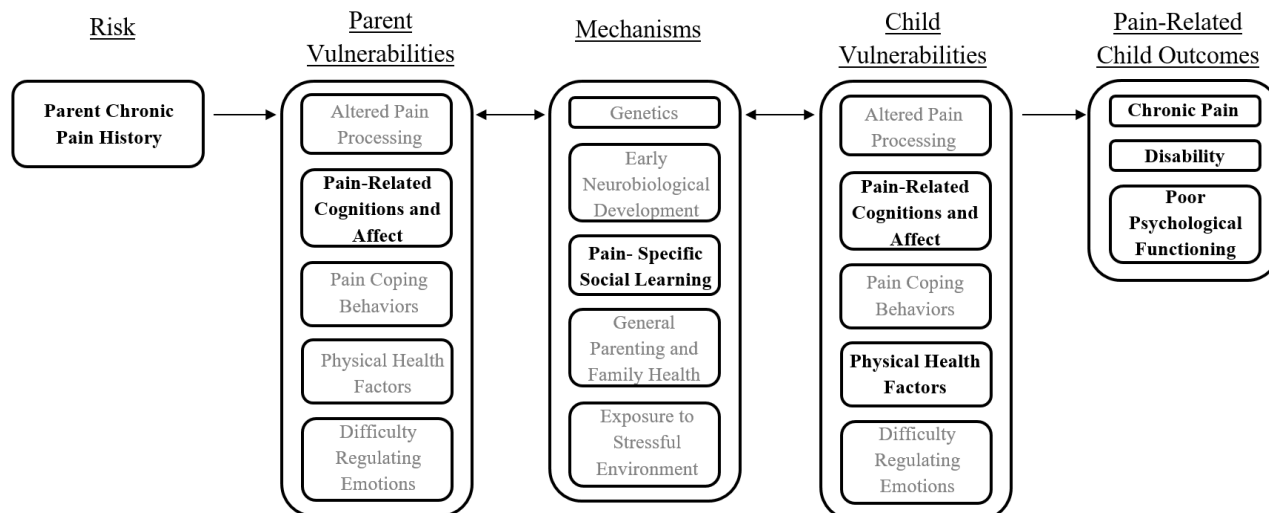


Figure 2. Conceptual Model of the Intergenerational Transmission of Parent Pain-Related Risk to Children's hEDS Experience (adapted from Stone & Wilson, 2016).

Parent Chronic Pain History

Many adults with hEDS are likely parents of children with hEDS (De Wandele et al., 2013) and their own history of chronic pain likely plays a role in their relationship with their children. Mothers with hEDS report that pain and fatigue together contribute to parental limitations, which are associated with feelings of inadequate parenting (De Baets et al., 2017). Parents may also suffer from depleted cognitive resources, related to expending resources towards coping with their own pain condition in addition to their regular responsibilities, potentially leading to inconsistent expressions of warmth, affection, discipline, withdrawal, anger, and irritability (Evans & de Souza, 2008; Umberger, Risko, & Covington, 2015; Wilson & Fales, 2015). Parents with chronic pain commonly report that their parenting has been negatively impacted in multiple ways due to their chronic pain condition (Wilson & Fales, 2015), which may indirectly affect child psychosocial functioning (Chen, 2017). Parent pain experience and functioning are also important predictors of child outcomes. Parent pain is predictive of adolescent's pain frequency, pain intensity, somatic symptoms, and pain-related disability

(Wilson et al., 2014). Parents with higher levels of pain interference with activities, are also more likely to have children with higher levels of current pain (Schanberg et al., 2001).

In thinking about where differences lie between parents with and without chronic pain, parents with chronic pain may think about pain and respond to pain differently than parents without chronic pain. Wilson and Fales (Wilson & Fales, 2015) suggest that parents with chronic pain may be more likely to catastrophize about their child's pain and respond to their child's pain with protective behaviors than parents without chronic pain. Wilson and colleagues have found that parents with more pain locations had higher parental catastrophizing about their adolescent's pain, concluding that parents who experience pain themselves are more likely to catastrophize about their adolescent's pain (Wilson et al., 2014). Wilson and Fales also found that parent's own activity interference due to pain, for parents with chronic pain, but not for those without, was associated with increased protective parenting (Wilson & Fales, 2015). Additionally, parents with chronic pain rated their adolescents as having more frequent and more intense pain than parents without chronic pain. When parents perceived their child was having more frequent and intense pain, parents with chronic pain responded with more protective response behaviors, while parents without chronic pain did not. Perhaps parent's own experiences with pain make them more attuned to picking up on their child's pain cues, but also may increase their risk of catastrophizing about their child's pain and may increase the likelihood that they will engage in protective behaviors in response to their child's pain (Wilson & Fales, 2015).

Parental Pain-Related Cognitions

Catastrophizing about pain involves thoughts, fears, and worries relating to an exaggerated negative perception of pain. Pain catastrophizing is characterized by the magnification of the impact of pain, readily evoked and prolonged rumination about pain, and

feeling helpless and unable to endure or stop the pain (Quartana, Campbell, & Edwards, 2009). Pain catastrophizing exerts influence on pain-related outcomes through multiple mechanisms. Pain catastrophizing leads to increased attention toward pain, (i.e. selectively attending towards pain, thinking about pain more, and increasing vigilance to bodily sensations in anticipation of pain) (Edwards, Bingham, Bathon, & Haythornthwaite, 2006). Catastrophizing also represents a negative appraisal of the pain experience and is associated with feelings of hopelessness and reduced self-efficacy (Edwards et al., 2006). Together, these beliefs interfere with engaging in active pain coping and beneficial health behaviors like exercise (Edwards et al., 2006; Quartana et al., 2009). Additionally, expressions of catastrophizing elicit solicitous responses to pain (Buenaver, Edwards, & Haythornthwaite, 2007), which help to reinforce and thus increase pain behaviors (Lynch-Jordan, Kashikar-Zuck, Szabova, & Goldschneider, 2013) and catastrophizing beliefs. Pain catastrophizing additionally has a direct effect on pain processing by altering endogenous pain modulation pathways of the central nervous system by promoting sensitization to pain and/ or interfering with pain inhibition (Edwards et al., 2006; Quartana et al., 2009). Pain catastrophizing and perception of pain are therefore intrinsically linked.

Consequently, how parents think about their own pain impacts and is impacted by their pain experiences. Parent's tendency to catastrophize about their own pain is positively related to their tendency to catastrophize about their child's pain (Goubert, Vervoort, Sullivan, Verhoeven, & Crombez, 2008). Parental catastrophizing about their own pain (Langer, Romano, Levy, Walker, & Whitehead, 2009; Wilson & Fales, 2015) and child's pain influences how parents respond to their children's expressions of pain (Hechlerl et al., 2011; Jaaniste et al., 2016; Langer, Romano, Mancl, & Levy, 2014). Therefore, parental catastrophizing about their child's

pain can be considered a parent vulnerability that influences the mechanism of pain-specific social learning.

Pain-Specific Social Learning

Social learning theory accounts for a major source of parental influence on child illness behaviors (i.e. observable expressions of child's pain or other symptomology). Two sources of social learning are parent modeling of how they cope with pain and parent response to their child's illness behaviors (Palermo & Chambers, 2005; Stone & Wilson, 2016; Walker & Zeman, 1992). Parents teach their children how to respond to pain symptoms by modeling their own illness behaviors and beliefs (Levy, 2010; Walker & Zeman, 1992). For instance, when parents stay at home from work or expect special privileges (i.e. choosing the family night movie, or reduction in household chores) when they are in pain, they model these behaviors and expectations for their children.

Similarly, parents teach their children what pain means to them through the way they respond to their child's expressions of pain. Frequently, parents will reinforce child illness behaviors. Expressions of support, care, concern, and attempts to make things easier for their child by allowing their child to skip chores, delay homework, and miss school, increase positive consequences of illness, promoting future pain expression (Levy, 2010). These types of parental response behaviors are referred to as protective or solicitous behaviors. Another consequence of rewarding children's symptomatic complaints is that children begin to attend to their symptoms more and can become sensitized to pick up on lower thresholds of pain (Levy, 2010; Walker, Garber, & Greene, 1991). Research has shown that these protective and solicitous parental behaviors are related to increased pain and disability in children with chronic pain (Claar, Simons, & Logan, 2008; Langer et al., 2009; Levy, 2010).

Some parents consciously adapt their behavior to reflect the message they wish to convey. For parents with their own chronic pain, this may have a greater significance as there are more opportunities for them to model their own illness behavior. For instance, some mothers with hEDS recognize their “double role” as a role model for their children, not wanting to express that pain needs to be endured, but also being a positive role model by showing what is possible for their children’s futures despite having this medical condition (De Baets et al., 2017). One mother gave the example of going to work when she is in pain to show her child that they cannot stay home from school every time they are not feeling well (De Baets et al., 2017).

Child Pain-Related Cognitions

Child pain catastrophizing is associated with greater pain intensity, depressive symptoms, anxiety symptoms, functional disability, and reduced quality of life, with higher catastrophizing predicting worse outcomes (Langer et al., 2009; Lynch-Jordan et al., 2013; Pielech et al., 2014). Due to negative outcomes associated with higher pain-catastrophizing, understanding how children come to develop catastrophizing beliefs is incredibly important. Development of child pain catastrophizing has genetic and social learning roots, both of which parents play a role in.

Parent’s own pain catastrophizing beliefs are related to children’s pain catastrophizing beliefs and pain-related outcomes. Trost and colleagues found that pain catastrophizing beliefs are 37% heritable (Trost et al., 2015). Therefore, children with parents who hold pain catastrophizing beliefs may be predisposed toward catastrophizing (Trost et al., 2015).

Another way parent’s own pain catastrophizing beliefs influence children’s pain catastrophizing beliefs is through protective response behaviors (Langer et al., 2009; Wilson & Fales, 2015). It has been found that the way parents respond to their child’s pain behaviors depends on their beliefs about pain (Langer et al., 2009; Langer et al., 2014). For example,

parents who have more catastrophizing thoughts about their own pain (Langer et al., 2009; Wilson & Fales, 2015) or their child's pain (Caes, Vervoort, Eccleston, Vandenhende, & Goubert, 2011) have reported exhibiting more protective response behaviors to their child's somatic (Langer et al., 2009) and pain (Langer et al., 2009; Wilson & Fales, 2015) complaints. Therefore, while child pain behavior expression predicted protective parental responding, parental pain catastrophizing mediated the effect (Langer et al., 2014). Parent reported protective response behaviors have also been found to mediate the influence of parental catastrophizing about their own pain on child functional disability (Langer et al., 2009). Together, this provides evidence for a parental pain catastrophizing - protective parental responding - child outcomes pathway.

Parent's pain catastrophizing has also been found to influence child outcomes through impacting child pain catastrophizing (Pielech et al., 2014; Wilson et al., 2014). While high catastrophizing parents are likely to have adolescents who have significantly more depressive symptoms, greater functional disability, higher pain intensity and more pain behaviors (Lynch-Jordan et al., 2013; Pielech et al., 2014), child and adolescent pain catastrophizing mediates the relationship between parental pain catastrophizing and child psychological and physical outcomes (Pielech et al., 2014; Wilson et al., 2014). Therefore, in cases in which parental pain catastrophizing has been found to influence child outcomes, the influence is indirect (Pielech et al., 2014; Wilson et al., 2014).

Completing the path, parental protective responses have been found to predict child functional disability, with child pain catastrophizing mediating this relationship (Cunningham et al., 2014; Guite, McCue, Sherker, Sherry, & Rose, 2011; Welkom, Hwang, & Guite, 2013). Welkom and colleagues (Welkom et al., 2013) concluded that parents who exhibit a greater

frequency of protective parenting responses tend to have children with stronger pain catastrophizing beliefs, which in turn, leads to increased functional disability in youth. Similarly, Cunningham and colleagues (Cunningham et al., 2014) speculated that parental protectiveness responses may increase catastrophic thinking in their children which may promote increased disability in youth.

When considered altogether, this series of studies suggests that in response to observing their child in pain, parents who exhibit higher levels of pain catastrophizing (about their own pain and/or their child's pain), respond with more protective behaviors. Protective behaviors teach and reinforce the child's catastrophic thinking about pain, which reduces effective coping with pain, leading to increased child pain intensity, functional disability, and psychological distress. Figure 3 demonstrates this proposed pathway.

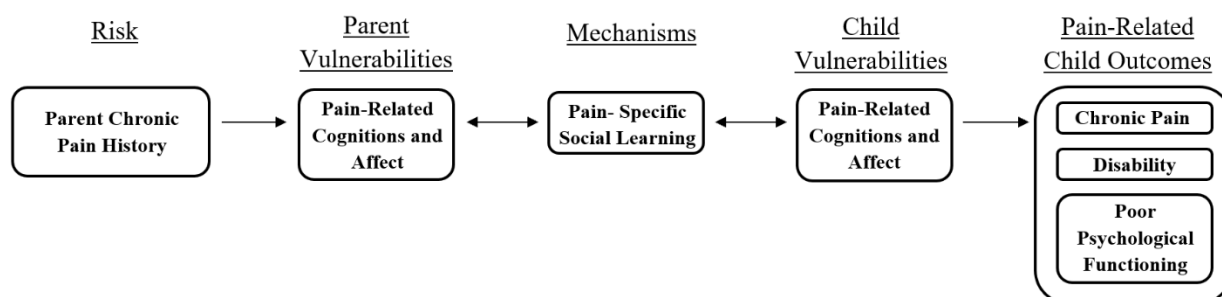


Figure 3. Variables from the Conceptual Model of the Intergenerational Transmission of Parent Pain-Related Risk to Children's hEDS Experience examined in the current study.

Rationale

It has been shown that children and adolescents with chronic pain including those with hEDS are at risk for negative psychosocial functioning and reduced HRQoL. The greater pediatric chronic pain and transgenerational pain risk literature draws attention to the multiple pathways through which parents influence their children's psychological and other pain-related outcomes, however, the relationship between parental hEDS experience and child hEDS

experience has yet to be explored. Due to the substantial impact that hEDS may have on a parent's psychological, physical functioning and parenting, it is possible that they think about pain and respond to their children's pain complaints differently than parents without a history of pain. Because it has been suggested that parents with chronic pain may be more likely to catastrophize about pain and pass these beliefs onto their children through increased use of protective response behaviors, parents with hEDS may put their children at additional risk for increased pain intensity, functional disability, anxiety, and depression. It is clinically important to identify any additional risks for worse pain-related child outcomes and the pathway through which the risk is transmitted in order to design targeted family treatment and prevention programs that minimize risk for families affected by hEDS.

Statement of Hypotheses

Hypothesis I. There will be a positive association between parent pain catastrophizing beliefs, protective parental response behaviors, child pain catastrophizing beliefs, child pain intensity, functional disability, anxiety symptoms, and depressive symptoms in children with hEDS.

Hypothesis II. Parents with hEDS or other chronic pain will have higher pain catastrophizing and report more protective response behaviors than parents with no history of chronic pain. There is limited literature on hEDS parent-child related factors, but the larger pediatric pain literature demonstrates that parents with varied chronic pain conditions may have this response to their children's pain. Thus, I expect parents with hEDS and other chronic pain to be similar to each other and dissimilar to parents without pain.

Hypothesis III. Children of parents with hEDS, other chronic pain, and without chronic pain will differ in pain-related outcomes: pain intensity, functional disability, symptoms of

anxiety and depression. Specifically, children of parents with hEDS and chronic pain will have worse pain-related outcomes than children of parents without chronic pain.

Hypothesis IV. Parental pain catastrophizing will predict child-pain related outcomes (pain intensity, functional disability, symptoms of anxiety, and symptoms of depression), through the effect that parent pain catastrophizing has on protective parental response behaviors.

Hypothesis V. Parental pain catastrophizing will predict child-pain related outcomes (pain intensity, functional disability, symptoms of anxiety and symptoms of depression) through the effect that parent pain catastrophizing has on child pain catastrophizing.

Hypothesis VI. Parental protective response behaviors will predict child-pain related outcomes (pain intensity, functional disability, symptoms of anxiety and symptoms of depression) through the effect that parental protective response behaviors have on child pain catastrophizing.

Supplemental Hypotheses:

Hypothesis VII. Parent pain catastrophizing about their child's pain will be strongly associated with child pain catastrophizing.

Method

Participants

Youth with hEDS and their parents will be recruited through two methods: in person during an hEDS clinic appointment within the Division of Clinical Genetics of Advocate Children's Hospital or online through a posting to an hEDS support group on Facebook. Patients approached at the clinic were screened for eligibility by the study Geneticist during their medical appointments. Patients who were diagnosed with hEDS using the Villefranche criteria (Beighton, Paepe, Steinmann, Tsiouras, & Wenstrup, 1998) were eligible to participate. The online survey

will include screening questions completed by both parents and youths to confirm a diagnosis of hEDS. Inclusion criteria additionally requires that youths be between 8 and 18 years old, speak and read English fluently. Participants recruited through the clinic were additionally screened for developmental delay and intellectual disability that would interfere with their ability to assent and answer study questionnaires. For families participating online, parents were directed to help explain the study to their child and complete the study questions with their child if their child had difficulty understanding.

Procedures

Clinic families who provided in person consent and assent were provided parent and child questionnaire packets to complete either in clinic or to mail back with pre-labeled and stamped envelopes. Both the Institutional Review Board (IRB) at DePaul University and Advocate Children's Hospital provided approval for the in-person portion of the study.

The organizer of the hEDS support group provided preliminary approval for a recruitment link to the study to be posted to the group's discussion board, contingent on appropriate IRB approval. Upon acceptance of this proposal, an application for the online recruitment of participants for the current study will be submitted to DePaul's IRB. We will request a "waiver of documentation of consent" from the IRB. The online survey will request parents and youths to select "I agree" or "I do not agree" in response to the consent and assent form. If "I do not agree" is selected, the survey will automatically discontinue. In order for children under the age of 18 to provide assent and participate, a parent or guardian will have to read and agree to the "parent permission for a child to participate in research" form first. Parents and youths will complete the same questionnaires as those administered to the clinic families, in electronic form.

Measures

Demographic information. The following demographic information was collected: the patient's age, sex, race, and ethnicity; the family's income, and the time since hEDS diagnosis.

Parent Measures

Parental hEDS and Chronic Pain Status. Parents reported family history of hEDS and chronic pain in response to open ended questions asking parents to report "other family member with EDS" and "other family members with chronic pain (of any type) and what type." Parents' pain status will be determined by comparing the relationship of the person completing the form to family members with EDS and pain.

Pain Catastrophizing Scale Parent Version (PCS-P). The PCS-P is a 13-item parent self-report that assess parent's catastrophic thinking about their child's pain on a 5-point scale in which 0 = *not at all*, 1 = *mildly*, 2 = *moderately*, 3 = *severely*, 4 = *extremely* (Goubert, Eccleston, Vervoort, Jordan, & Crombez, 2006). Items include "When my child is in pain..." "I keep thinking about how much I want the pain to stop," "I become afraid that the pain will get worse," and "it's awful and I feel that it takes over me." Scores range from 0 to 52 with higher scores indicating greater pain catastrophizing. These scores reflect multiple domains of pain catastrophizing including rumination, magnification and feelings of helplessness regarding pain (Crombez et al., 2003). Criterion validity and reliability of the PCS-P have been found in a sample of parents of adolescents with chronic pain (Goubert et al., 2006).

Adult Responses to Children's Symptoms (ARCS) Protect Subscale. The ARCS assesses parents' responses to their child's pain complaints (Van Slyke & Walker, 2006). The Protect scale of the ARCS includes behaviors in which the child receives special attention, treatment, privileges and reduced responsibility expectations (Van Slyke & Walker, 2006). The

scale contains 13-items in which caregivers are asked to indicate how often they engage in various behaviors using a 5-point Likert-type scale in which 0 = *never*, 1 = *once in a while*, 2 = *sometimes*, 3 = *often* 4 = *always*. Example items include “When you child is in pain, how often do you...” “Let your child sleep later than usual in the morning”, “Stay home from work or come home early (or stay home instead of going out or running errands),” and “Tell your child that he/she doesn’t have to finish his/her homework.” Scores range from 0 to 52 with higher scores indicating use of more protective response behaviors by the parent. This structure of the ARCS Protect Subscale is suggested for use with combined child and adolescent populations (Noel et al., 2015). The factor structure of the Protect subscale has been validated and strong reliability has been found in a sample of various pediatric chronic pain conditions and pain-related illnesses (Noel et al., 2015).

Child Measures

Pain Catastrophizing Scale Child Version (PCS-C). The PCS-C is a 13-item self-report measure that assesses children and adolescent’s catastrophic beliefs about their own pain experiences (Crombez et al., 2003). It assesses the same domains as the PCS-P, is on the same 5-point scale, and asks the same questions, but with a different item prompt: “When I am in pain...”. Scores range from 0 to 52 with higher scores indicating greater pain catastrophizing. The PCS-C has been validated for youth ages 8 to 16 with and without chronic pain (Crombez et al., 2003).

Pain Intensity. Children and adolescents reported their “usual level of pain in the last 2 weeks” on an 11-point numeric rating scale ranging from 0 = *No Pain at all* to 10 = *Worst Pain I Can Imagine*. This scale has been found to be a valid and reliable assessment of children’s pain intensity (Castarlenas, Jensen, von Baeyer, & Miró, 2017).

Patient Reported Outcomes Measurement Information System (PROMIS) Pediatric Anxiety Subscale. The Short Form Anxiety subscale is a part of the PROMIS Pediatric Scales and assesses a child's experience of anxious symptoms over a one-week period for patients between the ages of 8 and 17 years living with chronic illnesses (Varni et al., 2014). The Anxiety Short Form subscale contains 8 items. The measure asks children to report how often they have experienced different feelings over the past 7 days. Responses are reported using a 5-point Likert-type scale, where 0 = *Never*, 1 = *Almost Never*, 2 = *Sometimes*, 3 = *Often*, and 4 = *Almost Always*. Sample items on the Anxiety- Short Form subscale include, "I felt nervous," "I felt worried," and "I got scared really easy." Responses to items on each scale are summed to create an Anxiety Symptom subscale score ranging from 0-32. Raw scores from the short-form measure are converted to scaled T-scores (mean = 50). Higher T-scores indicate more anxious symptoms. It has been shown that the short form of the anxiety symptom subscale of the PROMIS measure is sufficient to provide precise measures of the symptoms (Irwin et al., 2010).

PROMIS Pediatric Depression Subscale. The Short Form Depression subscale is a part of the PROMIS Pediatric Scales and assesses a child's experience of depressive symptoms over a one-week period for patients between the ages of 8 and 17 years living with chronic illnesses (Varni et al., 2014). The Depression Short Form subscale contains 8 items. The measure asks children to report how often they have experienced different feelings over the past 7 days. Responses are reported using a 5-point Likert-type scale, where 0 = *Never*, 1 = *Almost Never*, 2 = *Sometimes*, 3 = *Often*, and 4 = *Almost Always*. Sample items on the Depressive Symptoms- Short Form subscale include, "I could not stop feeling sad," "I felt lonely," and "It was hard for me to have fun." Responses to items on each scale are summed to create a Depression Symptom subscale score ranging from 0-32. Raw scores from the short-form measure is converted to

scaled T-scores (mean = 50). Higher T-scores indicate more depressive symptoms. It has been shown that the short form of the depressive symptom subscale of the PROMIS measure is sufficient to provide precise measures of the symptoms (Irwin et al., 2010).

Functional Disability Inventory (FDI). The FDI measures “physical functioning and disability in youth with chronic pain” in the home, school, recreational, and social domains (Kashikar-Zuck et al., 2011, p. 1). The FDI contains 15 items with responses measured on a 5-point Likert-type scale in which 0 = *No Trouble*, 1 = *A Little Trouble*, 2 = *Some Trouble*, 3 = *A Lot of Trouble*, and 4 = *Impossible*. The measure asks youths to rate how much “physical trouble or difficulty” the child has doing each activity. Sample items include, “Walking up stairs,” “Eating regular meals,” “Reading or doing homework,” and “Getting to sleep at night and staying asleep.” Responses to items are summed to create total scores ranging from 0 to 60 with higher scores indicating greater pain-related disability. Children’s level of functional disability may be categorized as “No/Minimal Disability” (FDI score ≤ 12), “Moderate Disability” (FDI score 13 - 29), or “Severely Disabled” (FDI score ≥ 30) (Kashikar-Zuck et al., 2011). The FDI has been widely used with youth between the ages of 8 and 18 years (Kashikar-Zuck et al., 2011). Strong internal consistency, test-retest reliability, and parent-child concordance have been reported (Claar & Walker, 2006).

Analytic Plan

The current study will include data from about 100 children and adolescents between the ages of 8 and 18 years and a parent or caregiver of the child. Analyses will be conducted using SPSS Statistics and R Studio.

Hypothesis I proposes that there will be a positive association between protective parenting behaviors, parent pain catastrophizing beliefs, increased pain intensity, functional

disability, anxiety symptoms, depressive symptoms, and child pain catastrophizing beliefs, in children with hEDS. Bivariate correlations will be conducted to test the associations between the eight variables.

Hypothesis II proposes that parents with hEDS or other chronic pain will have higher pain catastrophizing and report more protective response behaviors than parents with no history of chronic pain. Two ANOVAs will be conducted to test whether there is a difference in pain catastrophizing scores and parent reported protective response behaviors between parents with hEDS, parents with other chronic pain, and parents without a history of chronic pain.

Hypothesis III proposes that children of parents with hEDS, with other chronic pain, and without chronic pain will differ in pain-related outcomes: pain intensity, functional disability, symptoms of anxiety and depression. Four ANOVAs will be conducted to test whether there is a difference in each of the four pain-related child outcomes between parents with hEDS, parents with other chronic pain and parents without a history of chronic pain.

Hypothesis IV proposes a set of four mediations (modeled in Figure 4) in which parent protective behaviors mediates the relationship between parental pain catastrophizing and child-pain related outcomes: pain intensity, functional disability, symptoms of anxiety, and symptoms of depression. Parental pain catastrophizing will be the predictor, protective parent response behaviors will serve as the mediator, and pain intensity, functional disability, anxiety symptoms, and depressive symptoms will be the outcome variables in each of the four mediation models.

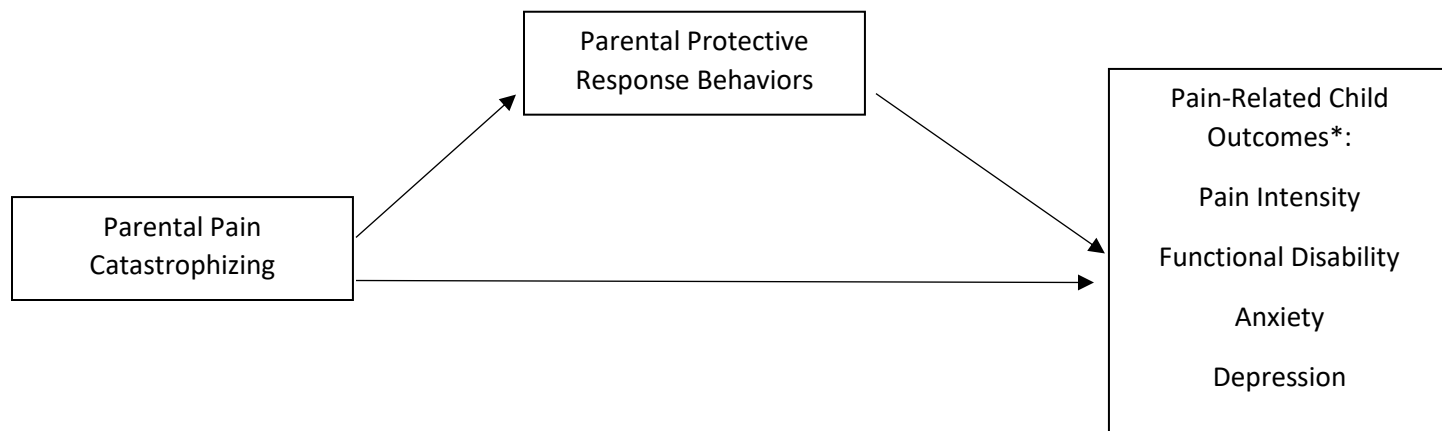


Figure 4. Mediation model of the effect of Parental Pain Catastrophizing on Pain-Related Child Outcomes, mediated by Parental Protective Response Behaviors. * Each outcome will be tested within its own mediation analysis: Pain Intensity, Functional Disability, Anxiety, Depression, respectively.

Hypothesis V proposes that a set of four mediations (modeled in Figure 5) in which child pain catastrophizing mediates the relationship between parental pain catastrophizing and child-pain related outcomes: pain intensity, functional disability, symptoms of anxiety and symptoms of depression. Parental pain catastrophizing will be the predictor, child pain catastrophizing will serve as the mediator, and pain intensity, functional disability, anxiety symptoms, and depressive symptoms will be the outcome variables in each mediation.

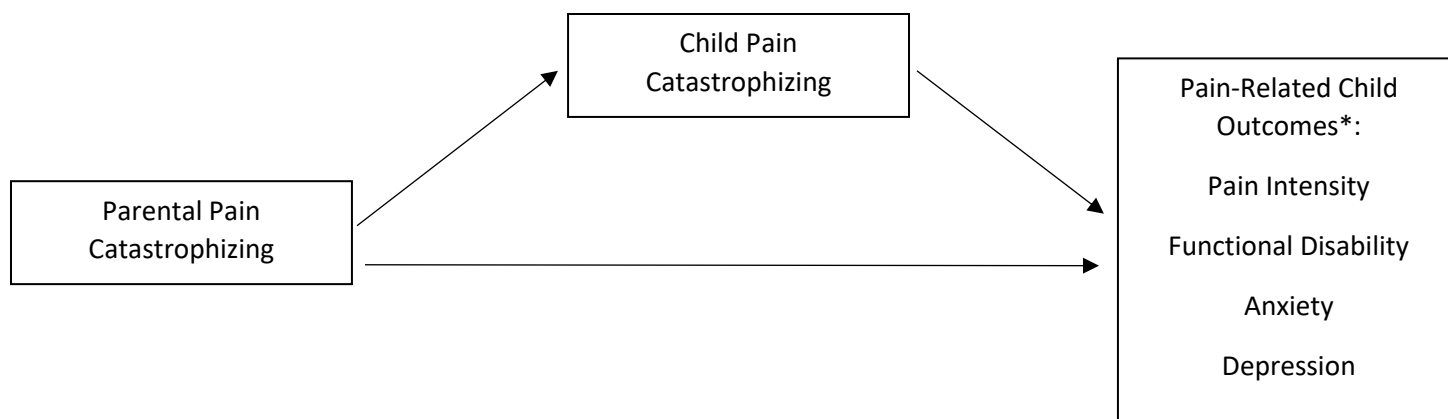


Figure 5. Mediation model of the effect of Parental Pain Catastrophizing on Pain-Related Child Outcomes, mediated by Child Pain Catastrophizing. * Each outcome will be tested within its own mediation analysis: Pain Intensity, Functional Disability, Anxiety, Depression, respectively.

Hypothesis V proposes a set of four mediations (modeled in Figure 6) in which child pain catastrophizing mediates the relationship between parental protective response behaviors and child-pain related outcomes: pain intensity, functional disability, symptoms of anxiety, and symptoms of depression. Parental protective response behaviors will be the predictor, child pain catastrophizing will serve as the mediator, and pain intensity, functional disability, anxiety symptoms, and depressive symptoms will be the outcome variables in each mediation.

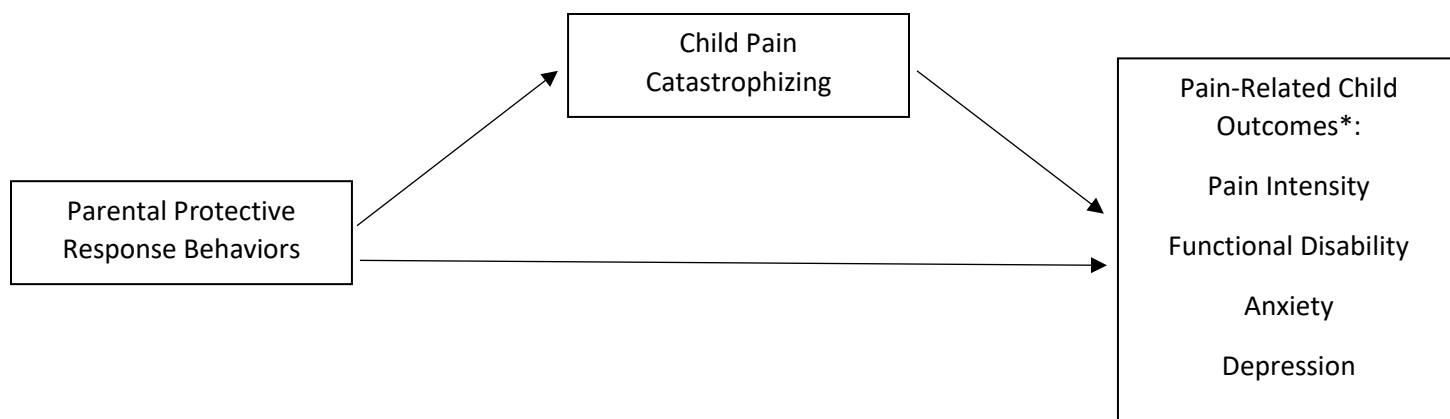


Figure 6. Mediation model of the effect of Parental Protective Response Behaviors on Pain-Related Child Outcomes, mediated by Child Pain Catastrophizing. * Each outcome will be tested within its own mediation analysis: Pain Intensity, Functional Disability, Anxiety, Depression, respectively.

Supplemental Analysis:

Hypothesis VI proposes that parent pain catastrophizing about their child's pain will correspond closely with/predict child pain catastrophizing beliefs. A paired samples t-test will be run to test whether each parent and their child report significantly different pain catastrophizing beliefs or not.

References

- Adib, N., Davies, K., Grahame, R., Woo, P., & Murray, K. (2005). Joint hypermobility syndrome in childhood. A not so benign multisystem disorder? *Rheumatology*, *44*(6), 744-750.
- Baeza-Velasco, C., Bourdon, C., Montalescot, L., de Cazotte, C., Pailhez, G., Bulbena, A., & Hamonet, C. (2018). Low-and high-anxious hypermobile Ehlers–Danlos syndrome patients: comparison of psychosocial and health variables. *Rheumatology international*, *38*(5), 871-878.
- Beighton, P., Paepe, A. D., Steinmann, B., Tsipouras, P., & Wenstrup, R. J. (1998). Ehlers-Danlos syndromes: revised nosology, Villefranche, 1997. *American journal of medical genetics*, *77*(1), 31-37.
- Buenaver, L. F., Edwards, R. R., & Haythornthwaite, J. A. (2007). Pain-related catastrophizing and perceived social responses: Inter-relationships in the context of chronic pain. *Pain*, *127*(3), 234-242.
- Bulbena, A., Baeza-Velasco, C., Bulbena-Cabré, A., Pailhez, G., Critchley, H., Chopra, P., . . . Porges, S. (2017). *Psychiatric and psychological aspects in the Ehlers–Danlos syndromes*. Paper presented at the American Journal of Medical Genetics Part C: Seminars in Medical Genetics.
- Caes, L., Vervoort, T., Eccleston, C., Vandenhende, M., & Goubert, L. (2011). Parental catastrophizing about child's pain and its relationship with activity restriction: the mediating role of parental distress. *PAIN®*, *152*(1), 212-222.

- Castarlenas, E., Jensen, M. P., von Baeyer, C. L., & Miró, J. (2017). Psychometric properties of the numerical rating scale to assess self-reported pain intensity in children and adolescents. *The Clinical journal of pain, 33*(4), 376-383.
- Castori, M., Dordoni, C., Valiante, M., Sperduti, I., Ritelli, M., Morlino, S., . . . Camerota, F. (2014). Nosology and inheritance pattern (s) of joint hypermobility syndrome and Ehlers-Danlos syndrome, hypermobility type: A study of intrafamilial and interfamilial variability in 23 Italian pedigrees. *American Journal of Medical Genetics Part A, 164*(12), 3010-3020.
- Castori, M., Morlino, S., Celletti, C., Celli, M., Morrone, A., Colombi, M., . . . Grammatico, P. (2012). Management of pain and fatigue in the joint hypermobility syndrome (aka Ehlers–Danlos syndrome, hypermobility type): principles and proposal for a multidisciplinary approach. *American Journal of Medical Genetics Part A, 158*(8), 2055-2070.
- Cattalini, M., Khubchandani, R., & Cimaz, R. (2015). When flexibility is not necessarily a virtue: a review of hypermobility syndromes and chronic or recurrent musculoskeletal pain in children. *Pediatric Rheumatology, 13*(1), 1.
- Chen, C. Y.-C. (2017). Effects of Parental Chronic Illness on Children's Psychosocial and Educational Functioning: a Literature Review. *Contemporary School Psychology, 21*(2), 166-176.
- Claar, R., Simons, L. E., & Logan, D. E. (2008). Parental response to children's pain: The moderating impact of children's emotional distress on symptoms and disability. *Pain, 138*(1), 172-179. doi:10.1016/j.pain.2007.12.005

- Claar, R., & Walker, L. (2006). Functional assessment of pediatric pain patients: psychometric properties of the functional disability inventory. *Pain, 121*(1), 77-84.
- Crombez, G., Bijttebier, P., Eccleston, C., Mascagni, T., Mertens, G., Goubert, L., & Verstraeten, K. (2003). The child version of the pain catastrophizing scale (PCS-C): A preliminary validation. *Pain, 104*(3), 639-646.
- Cunningham, N. R., Lynch-Jordan, A., Barnett, K., Peugh, J., Sil, S., Goldschneider, K., & Kashikar-Zuck, S. (2014). Child pain catastrophizing mediates the relationship between parent responses to pain and disability in youth with functional abdominal pain. *Journal of pediatric gastroenterology and nutrition, 59*(6), 732.
- De Baets, S., Vanhalst, M., Coussens, M., Rombaut, L., Malfait, F., Van Hove, G., . . . Van de Velde, D. (2017). The influence of Ehlers-Danlos syndrome–hypermobility type, on motherhood: A phenomenological, hermeneutical study. *Research in developmental disabilities, 60*, 135-144.
- De Wandele, I., Rombaut, L., Malfait, F., De Backer, T., De Paepe, A., & Calders, P. (2013). Clinical heterogeneity in patients with the hypermobility type of Ehlers-Danlos syndrome. *Research in developmental disabilities, 34*(3), 873-881.
- Denk, F., McMahon, S. B., & Tracey, I. (2014). Pain vulnerability: a neurobiological perspective. *Nature neuroscience, 17*(2), 192.
- Edwards, R. R., Bingham, C. O., Bathon, J., & Haythornthwaite, J. A. (2006). Catastrophizing and pain in arthritis, fibromyalgia, and other rheumatic diseases. *Arthritis Care & Research, 55*(2), 325-332.
- Engelbert, R. H., Juul-Kristensen, B., Pacey, V., De Wandele, I., Smeenk, S., Woinarosky, N., . . . Simmonds, J. V. (2017). *The evidence-based rationale for physical therapy treatment of*

- children, adolescents, and adults diagnosed with joint hypermobility syndrome/hypermobility Ehlers Danlos syndrome*. Paper presented at the American Journal of Medical Genetics Part C: Seminars in Medical Genetics.
- Evans, S., & de Souza, L. (2008). Dealing with chronic pain: giving voice to the experiences of mothers with chronic pain and their children. *Qualitative Health Research, 18*(4), 489-500.
- Fatoye, F., Palmer, S., Macmillan, F., Rowe, P., & van der Linden, M. (2012). Pain intensity and quality of life perception in children with hypermobility syndrome. *Rheumatology international, 32*(5), 1277-1284.
- Fikree, A., Chelimsky, G., Collins, H., Kovacic, K., & Aziz, Q. (2017). *Gastrointestinal involvement in the Ehlers–Danlos syndromes*. Paper presented at the American Journal of Medical Genetics Part C: Seminars in Medical Genetics.
- Forgeron, P. A., King, S., Stinson, J. N., McGrath, P. J., MacDonald, A. J., & Chambers, C. T. (2010). Social functioning and peer relationships in children and adolescents with chronic pain: A systematic review. *Pain Research and Management, 15*(1), 27-41.
- Garralda, E., & Rangel, L. (2004). Impairment and coping in children and adolescents with chronic fatigue syndrome: a comparative study with other paediatric disorders. *Journal of Child Psychology and Psychiatry, 45*(3), 543-552.
- Goubert, L., Eccleston, C., Vervoort, T., Jordan, A., & Crombez, G. (2006). Parental catastrophizing about their child's pain. The parent version of the Pain Catastrophizing Scale (PCS-P): a preliminary validation. *Pain, 123*(3), 254-263.

- Goubert, L., Vervoort, T., Sullivan, M. J., Verhoeven, K., & Crombez, G. (2008). Parental emotional responses to their child's pain: the role of dispositional empathy and catastrophizing about their child's pain. *The Journal of Pain, 9*(3), 272-279.
- Guite, J. W., McCue, R. L., Sherker, J. L., Sherry, D. D., & Rose, J. B. (2011). Relationships among pain, protective parental responses, and disability for adolescents with chronic musculoskeletal pain: the mediating role of pain catastrophizing. *The Clinical journal of pain, 27*(9), 775-781.
- Hakim, A., De Wandele, I., O'Callaghan, C., Pocinki, A., & Rowe, P. (2017). *Chronic fatigue in Ehlers–Danlos syndrome—Hypermobility type*. Paper presented at the American Journal of Medical Genetics Part C: Seminars in Medical Genetics.
- Hechlerl, T., Vervoortl, T., Hamannl, M., Tietzel, A. L., Vocksl, S., Goubertl, L., . . . Schroederl, S. (2011). Parental catastrophizing about their child's chronic pain: Are mothers and fathers different? *European Journal of Pain, 15*(5), 515. e511-515. e519.
- Hershenfeld, S. A., Wasim, S., McNiven, V., Parikh, M., Majewski, P., Faghfoury, H., & So, J. (2016). Psychiatric disorders in Ehlers–Danlos syndrome are frequent, diverse and strongly associated with pain. *Rheumatology international, 36*(3), 341-348.
- Irwin, D. E., Stucky, B., Langer, M. M., Thissen, D., DeWitt, E. M., Lai, J.-S., . . . DeWalt, D. A. (2010). An item response analysis of the pediatric PROMIS anxiety and depressive symptoms scales. *Quality of Life Research, 19*(4), 595-607.
- Jaaniste, T., Jia, N., Lang, T., Goodison-Farnsworth, E., McCormick, M., & Anderson, D. (2016). The relationship between parental attitudes and behaviours in the context of paediatric chronic pain. *Child: care, health and development, 42*(3), 433-438.

- Kashikar-Zuck, S., Flowers, S. R., Claar, R. L., Guite, J. W., Logan, D. E., Lynch-Jordan, A. M., . . . Wilson, A. C. (2011). Clinical utility and validity of the Functional Disability Inventory (FDI) among a multicenter sample of youth with chronic pain. *Pain, 152*(7), 1600-1607.
- Kashikar-Zuck, S., Goldschneider, K., Powers, S., Vaught, M., & Hershey, A. (2001). Depression and functional disability in chronic pediatric pain. *The Clinical journal of pain, 17*(4), 341-349.
- Kashikar-Zuck, S., Parkins, I. S., Graham, T. B., Lynch, A. M., Passo, M., Johnston, M., . . . Richards, M. M. (2008). Anxiety, mood, and behavioral disorders among pediatric patients with juvenile fibromyalgia syndrome. *The Clinical journal of pain, 24*(7), 620-626. doi:10.1097/AJP.0b013e31816d7d23
- Krahe, A. M., Adams, R. D., & Nicholson, L. L. (2018). Features that exacerbate fatigue severity in joint hypermobility syndrome/Ehlers–Danlos syndrome–hypermobility type. *Disability and rehabilitation, 40*(17), 1989-1996.
- Langer, S. L., Romano, J. M., Levy, R. L., Walker, L. S., & Whitehead, W. E. (2009). Catastrophizing and Parental Response to Child Symptom Complaints. *Children's Health Care, 38*(3), 169-184. doi:10.1080/02739610903038750
- Langer, S. L., Romano, J. M., Mancl, L., & Levy, R. L. (2014). Parental catastrophizing partially mediates the association between parent-reported child pain behavior and parental protective responses. *Pain research and treatment, 2014*. doi:10.1155/2014/751097
- Levy, R. L. (2010). Exploring the intergenerational transmission of illness behavior: from observations to experimental intervention. *Annals of Behavioral Medicine, 41*(2), 174-182.

- Logan, D. E., Simons, L. E., Stein, M. J., & Chastain, L. (2008). School impairment in adolescents with chronic pain. *The Journal of Pain, 9*(5), 407-416.
- Lynch-Jordan, A. M., Kashikar-Zuck, S., Szabova, A., & Goldschneider, K. R. (2013). The interplay of parent and adolescent catastrophizing and its impact on adolescents' pain, functioning, and pain behavior. *The Clinical journal of pain, 29*(8), 681.
- Malfait, F., Francomano, C., Byers, P., Belmont, J., Berglund, B., Black, J., . . . Burrows, N. P. (2017). *The 2017 international classification of the Ehlers–Danlos syndromes*. Paper presented at the American Journal of Medical Genetics Part C: Seminars in Medical Genetics.
- Mulvey, M. R., Macfarlane, G. J., Beasley, M., Symmons, D. P., Lovell, K., Keeley, P., . . . McBeth, J. (2013). Modest Association of Joint Hypermobility With Disabling and Limiting Musculoskeletal Pain: Results From a Large-Scale General Population–Based Survey. *Arthritis Care & Research, 65*(8), 1325-1333.
- Noel, M., Palermo, T. M., Essner, B., Zhou, C., Levy, R. L., Langer, S. L., . . . Walker, L. S. (2015). A developmental analysis of the factorial validity of the parent-report version of the Adult Responses to Children's Symptoms in children versus adolescents with chronic pain or pain-related chronic illness. *The Journal of Pain, 16*(1), 31-41.
- Pacey, V., Adams, R. D., Tofts, L., Munns, C. F., & Nicholson, L. L. (2014). Joint hypermobility syndrome subclassification in paediatrics: a factor analytic approach. *Archives of disease in childhood, archdischild-2013-305304*.
- Pacey, V., Tofts, L., Adams, R. D., Munns, C. F., & Nicholson, L. L. (2013). Exercise in children with joint hypermobility syndrome and knee pain: a randomised controlled trial

- comparing exercise into hypermobile versus neutral knee extension. *Pediatric Rheumatology*, *11*(1), 30.
- Pacey, V., Tofts, L., Adams, R. D., Munns, C. F., & Nicholson, L. L. (2015). Quality of life prediction in children with joint hypermobility syndrome. *Journal of paediatrics and child health*, *51*(7), 689-695.
- Palermo, T. M., & Chambers, C. T. (2005). Parent and family factors in pediatric chronic pain and disability: an integrative approach. *Pain*, *119*(1-3), 1-4.
doi:10.1016/j.pain.2005.10.027
- Pielech, M., Ryan, M., Logan, D., Kaczynski, K., White, M. T., & Simons, L. E. (2014). Pain catastrophizing in children with chronic pain and their parents: Proposed clinical reference points and reexamination of the Pain Catastrophizing Scale measure. *PAIN®*, *155*(11), 2360-2367.
- Quartana, P. J., Campbell, C. M., & Edwards, R. R. (2009). Pain catastrophizing: a critical review. *Expert review of neurotherapeutics*, *9*(5), 745-758.
- Rombaut, L., Malfait, F., De Paepe, A., Rimbaut, S., Verbruggen, G., De Wandele, I., & Calders, P. (2011). Impairment and impact of pain in female patients with Ehlers-Danlos syndrome: A comparative study with fibromyalgia and rheumatoid arthritis. *Arthritis & Rheumatism*, *63*(7), 1979-1987.
- Schanberg, L. E., Anthony, K. K., Gil, K. M., Lefebvre, J. C., Kredich, D. W., & Macharoni, L. M. (2001). Family pain history predicts child health status in children with chronic rheumatic disease. *Pediatrics*, *108*(3), e47-e47.
- Scheper, M. C., Juul-Kristensen, B., Rombaut, L., Rameckers, E. A., Verbunt, J., & Engelbert, R. H. (2016). Disability in adolescents and adults diagnosed with hypermobility-related

- disorders: a meta-analysis. *Archives of physical medicine and rehabilitation*, 97(12), 2174-2187.
- Schubert-Hjalmarsson, E., Öhman, A., Kyllerman, M., & Beckung, E. (2012). Pain, balance, activity, and participation in children with hypermobility syndrome. *Pediatric Physical Therapy*, 24(4), 339-344.
- Sinibaldi, L., Ursini, G., & Castori, M. (2015). *Psychopathological manifestations of joint hypermobility and joint hypermobility syndrome/Ehlers–Danlos syndrome, hypermobility type: The link between connective tissue and psychological distress revised*. Paper presented at the American Journal of Medical Genetics Part C: Seminars in Medical Genetics.
- Stone, A. L., & Wilson, A. C. (2016). Transmission of risk from parents with chronic pain to offspring: an integrative conceptual model. *Pain*, 157(12), 2628.
- Tinkle, B., Bird, H., Grahame, R., Lavalley, M., Levy, H., & Silience, D. (2009). The lack of clinical distinction between the hypermobility type of Ehlers–Danlos syndrome and the joint hypermobility syndrome (aka hypermobility syndrome). *American Journal of Medical Genetics Part A*, 149(11), 2368-2370.
- Tinkle, B., Castori, M., Berglund, B., Cohen, H., Grahame, R., Kazkaz, H., & Levy, H. (2017). *Hypermobility Ehlers–Danlos syndrome (aka Ehlers–Danlos syndrome Type III and Ehlers–Danlos syndrome hypermobility type): Clinical description and natural history*. Paper presented at the American Journal of Medical Genetics Part C: Seminars in Medical Genetics.

- Trost, Z., Strachan, E., Sullivan, M., Vervoort, T., Avery, A. R., & Afari, N. (2015). Heritability of pain catastrophizing and associations with experimental pain outcomes: a twin study. *Pain, 156*(3), 514.
- Umberger, W. A., Risko, J., & Covington, E. (2015). The forgotten ones: Challenges and needs of children living with disabling parental chronic pain. *Journal of pediatric nursing, 30*(3), 498-507.
- Van Slyke, D. A., & Walker, L. S. (2006). Mothers' responses to children's pain. *The Clinical journal of pain, 22*(4), 387-391.
- Varni, J. W., Magnus, B., Stucky, B. D., Liu, Y., Quinn, H., Thissen, D., . . . DeWalt, D. A. (2014). Psychometric properties of the PROMIS® pediatric scales: precision, stability, and comparison of different scoring and administration options. *Quality of Life Research, 23*(4), 1233-1243.
- Walker, L. S., Garber, J., & Greene, J. W. (1991). Somatization symptoms in pediatric abdominal pain patients: relation to chronicity of abdominal pain and parent somatization. *Journal of abnormal child psychology, 19*(4), 379-394.
- Walker, L. S., & Zeman, J. L. (1992). Parental response to child illness behavior. *Journal of pediatric psychology, 17*(1), 49-71.
- Welkom, J. S., Hwang, W.-T., & Guite, J. W. (2013). Adolescent pain catastrophizing mediates the relationship between protective parental responses to pain and disability over time. *Journal of pediatric psychology, jst011*.
- Wilson, A., & Fales, J. (2015). Parenting in the context of chronic pain: a controlled study of parents with chronic pain. *The Clinical journal of pain, 31*(8), 689.

Wilson, A., Moss, A., Palermo, T., & Fales, J. (2014). Parent pain and catastrophizing are associated with pain, somatic symptoms, and pain-related disability among early adolescents. *Journal of pediatric psychology, 39*(4), 418-426.

Appendix C. Additional Results

In the original proposal, Hypothesis VI proposed that parent pain catastrophizing about their child's pain would correspond closely with/predict child pain catastrophizing beliefs. A paired samples t-test was planned to test whether each parent and their child report significantly differed in pain catastrophizing beliefs or not. The results of the paired samples t-test were not significant indicating that parents and children's pain catastrophizing beliefs were not significantly different from each other. Therefore, within families, parents and children had similar levels of pain catastrophizing beliefs.