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The Role of Functional Disability and Social Support in Psychological Outcomes for Individuals with Pediatric Hypermobile Ehlers-Danlos Syndrome

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**The Role of Functional Disability and Social Support in Psychological Outcomes for
Individuals with Pediatric Hypermobile Ehlers-Danlos Syndrome**

A Thesis

Presented in

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

By

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May 2021

Department of Psychology
College of Science and Health
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Thesis Committee

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Biography

The author was born in Geneva, IL, on December 15, 1996. Keely graduated from St. Charles East High School, in St. Charles, IL in 2015. She then received her BS in psychology, with minors in neuroscience and wellness, from North Central College in 2019.

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Abstract

Objectives: Individuals with Hypermobile Ehlers-Danlos Syndrome (hEDS), a genetic condition that impacts a person's connective tissues, report a large number of physical symptoms including chronic pain, fatigue, gastrointestinal dysfunction, and joint dislocations. Research into the psychosocial outcomes of this condition remains insufficient, especially for pediatric patients. The present study aims to fill this research gap by examining the relationship between functional disability, social support, and mental health outcomes for individuals with pediatric hEDS. We hypothesize that increased functional disability will be associated with increased mental health challenges, specifically anxiety and depression. Furthermore, it is hypothesized that this relationship will be moderated by general social support such that higher levels of social support will mitigate the negative psychological impacts of functional disability. The influence of gender on experiences with pediatric hEDS is also explored. **Methods:** Thirty-four youth with pediatric hEDS recruited from a Midwest genetics clinic completed self-report questionnaires regarding their functional disability, perceived social support, and mental health. **Results:** Results demonstrate associations between functional disability and mental health and social support and mental health independently. Social support was not found to moderate the relationship between functional disability and mental health. **Conclusions:** Functional disability and social support each have a unique influence on the mental health of children with pediatric hEDS. Exploratory analyses into the influence of gender provide a groundwork for future studies.

Key words: Hypermobile Ehlers-Danlos Syndrome, Mental Health, Social Support, Functional Disability

The Role of Functional Disability and Social Support in Psychological Outcomes for Individuals with Pediatric Hypermobile Ehlers-Danlos Syndrome

The present study focuses on pediatric Hypermobile Ehlers-Danlos Syndrome (hEDS), which is a subtype of a broader collection of genetic conditions called Ehlers-Danlos Syndrome (EDS). Broadly speaking, EDS impacts a person's connective tissues such as skin and joints (Malfait et al., 2017). Symptoms vary between subtypes, with some experiencing more joint laxity and others having an increased risk of cardiac complications (Malfait et al., 2017). hEDS is the most common of the 13 subtypes of EDS (U.S. National Library of Medicine, 2020). The hypermobility of joints associated with this subtype frequently results in chronic pain (CP; Malfait et al., 2017). The prevalence rate of hEDS is not clear, with some research suggesting somewhere between 1 in 5,000 and 1 in 20,000 people having the syndrome (Levy, 2018) and others suggesting that it may be more frequent (Tinkle, et al, 2017). The CP that occurs with hEDS can result in functional disability, which is difficulty in completing tasks of daily living, that can hinder other aspects of the individual's life (Voermans et al., 2010; Walker & Greene, 1991).

Aside from functional disability, social challenges, such as difficulty connecting with and being understood by peers, are observed in youth with hEDS and CP (Bieniak et al., Revise & Resubmit; Kashikar-Zuck et al., 2008). Psychological distress, such as anxiety and depression, is of concern for adults with hEDS (Murray et al., 2013) and this distress is observed at higher rates in those with the condition than in those without (Cederlöf et al., 2016). At present, there is minimal research in regard to the prevalence of psychological distress in pediatric hEDS; however, similar pediatric CP conditions are marked by increased risks of mental health challenges (Pavlova et al., 2017). Thus, it is likely that similar trends are present in youth with

pediatric hEDS. Thus, it is important to examine the psychological outcomes of the pediatric hEDS population and begin to understand the relationship between psychological wellbeing and other variables.

hEDS

EDS is thought to be the result of a genetic mutation that influences the creation of collagen within the body (National Institutes of Health, 2019; Tinkle, et al., 2017). Collagen is a protein that makes up fibers in an individual's body, and these fibers exist in joints, skin, and other connective tissues. Collagen fibers are what allow tissue to stretch and then return to their original state without damage (Lodish et al., 2000). While the reason for this genetic mutation is not yet understood, the physical impacts can be significant, as is seen in the CP and additional symptoms of hEDS.

Those living with hEDS experience a broad range of symptoms that vary in severity and frequency. The most typical physical symptoms include pain, fatigue, joint dislocations, and bruising of the skin (Tinkle, et al., 2017). Pain in hEDS is generally rated as more severe than those with the classical type of EDS and may be experienced as widespread pain or localized to a specific part of an individual's body (Voermans et al., 2010). The pain typically becomes more widespread throughout an individual's body with age (Syx et al., 2017). The CP that exists with hEDS occurs as a consequence of the dysfunctional connective tissue, but also as a consequence of injuries and dislocations that result from the dysfunctional connective tissue (Syx et al., 2017). Other physical manifestations of hEDS include gastrointestinal dysfunction, cardiovascular issues, postural orthostatic tachycardia syndrome, headaches and migraines, sleep disturbances, and dysautonomia (Tinkle, et al, 2017). The array of potential symptoms, different bodily

locations of pain, and spectrum of pain severity and frequency make the physical impacts of hEDS complex and unique to each individual with the condition.

As there is limited research on pediatric hEDS, the research on other CP conditions is utilized to predict how pain and functional disability will impact those with hEDS. Chronic pain, in hEDS and more generally, is a threat to the health and wellness of individuals across the world. CP is characterized as persistent, intrusive physical discomfort over a period of time longer than to be expected for an injury or acute condition (National Institutes of Health, 2019). In general, females tend to experience more chronic pain conditions, including hEDS, than males (King et al., 2011; Cederlöf et al., 2016), and females are more likely to be diagnosed with hEDS than males (Castori et al., 2010). Recent estimates suggest that approximately 20% of adults in the United States have experienced CP in their lifetime (Dahlhamer et al., 2016). Aside from physical consequences, in adult populations, CP results in decreased functional abilities, loss of productivity, and a high cost of care (Dahlhamer et al., 2016). There is substantial evidence supporting that CP is highly associated with depression (Fishbain et al., 1997), demonstrating that the physical symptoms of such conditions impact more than a person's physical wellbeing. CP does not only impact adults however, with estimates suggesting that between 20 and 40% of children experience chronic pain worldwide (Stanford et al., 2008; Friedrichsdorf et al., 2016).

Chronic pain conditions are complex, with patient quality of life being influenced by a variety of factors including pain, functional changes, and social support. Understanding how various components of an individual's life influence their CP can assist practitioners in providing care that gives attention to more than just the physical manifestations of the disorder.

hEDS and Functional Disability

The consequences of hEDS expand beyond physical symptoms and into other realms of wellbeing, including functioning and social support. Pediatric CP disorders are frequently marked by increased functional disability, including difficulty with daily tasks, activities, and the ability to care for oneself as a result of the disorder (Kashikar-Zuck et al., 2008; Palermo, & Kiska, 2005). A pattern of functional disability has been documented in pediatric hEDS patients as well (Voermans et al., 2010). The presence of common symptoms such as pain, fatigue, and gastrointestinal dysfunction negatively influence a person's ability to engage in typical daily tasks such as school, tasks requiring physical activity, and caring for oneself (Murray et al., 2013). For young children, this will impact typical patterns of play, engagement in school, and extracurricular activities such as athletics.

Functional disability has a particularly strong, positive relationship with pain intensity (Voermans et al., 2010; Simons et al., 2012) and this is important given that pain is the most commonly reported difficulty with having hEDS (Tran et al, 2020; Bieniak Revise & Resubmit). This relationship suggests that for pediatric patients with hEDS, as pain symptoms worsen, functional disability related to school, playing at recess, or walking up and down stairs at home will also increase. Further, as pain is associated with more psychological challenges (Hunfeld et al., 2001), the CP in hEDS not only impacts a person's physical wellbeing, but also their emotional wellbeing. Additionally, symptoms of psychological disorders are positively associated with increased levels of EDS pain (Hershenfeld et al., 2016). Thus, pain in hEDS has widespread physical and emotional consequences.

hEDS and Mental Health

Mental health outcomes are measured within this study by assessing the symptoms of anxiety and depression present in participants. According to the DSM-V, anxiety disorders are characterized by excessive worry that is difficult to control and impairs a person's functioning (American Psychiatric Association, 2013). Depression, under the DSM-V, is marked by depressed mood, loss of interest in activities that previously brought someone pleasure, feelings of hopelessness or worthlessness, sleep changes, unexplained weight loss or gain, and other symptoms that impact an individual's ability to function (American Psychiatric Association, 2013). Individuals with pediatric CP experience worse psychological outcomes than those without CP (Vinall et al., 2016; Pavlova et al., 2017) and this association exists across age of individual and type of CP condition (Fishbain et al., 1997).

Research on adults with both classic and hypermobile type EDS has consistently found an association between the condition and mental health disorders (Hershenfeld et al., 2016; Bulbena et al., 2012; Cederlöf et al., 2016; Pasquini et al., 2014). Anxiety and depression are the two most frequent psychological challenges faced by adults with EDS, with anxiety being slightly more prevalent than depression (Bulbena et al., 2017). Furthermore, in a study of adults with hEDS, 69% of participants self-reported experiencing depression and 73% reported experiencing anxiety (Murray et al., 2013). In comparison to healthy peers without hEDS, those with the condition have repeatedly been found to have higher rates of mental health challenges (Mu et al., 2019; Cederlöf et al., 2016). The strong association between mental health challenges and EDS (Hershenfeld et al., 2016) emphasizes how the syndrome affects not only physical wellbeing but also psychological wellness. Given these problems in adults with hEDS and

children with other CP conditions, it is crucial that this research gap be filled to improve the understanding of how various pediatric hEDS related factors influence mental health outcomes.

While there is little research into the mental health of children and adolescents with hEDS, general pediatric CP research has demonstrated high rates of comorbidity of CP and psychological disorders (Vinall et al., 2016). In a study of general CP in adolescents, about a quarter of participants had a co-occurring CP condition and a mental health condition (Tegethoff et al., 2015). It is important to note, however, that this study found that a significant number of these individuals had mental health disorders that preceded chronic pain, suggesting that more research is needed on the directionality of the relationship between CP and psychological outcomes. Additionally, some gender differences exist in the relationship between anxiety and CP, with adolescent females tending to report more anxiety in relation to CP than males (Pavlova et al., 2017).

hEDS and Social Support

Perceived social support is the extent to which an individual feels they are receiving support from individuals in their life (Ridenour et al., 2006). Social support can be assessed in a global sense, looking at a person's overall level of support, or in terms of specific domains or sources, such as parents, friends, classmates, and teachers (Ridenour et al., 2006). There is significant variability in what individuals want their social support to look like. For some, social support may take the form of words of encouragement, accommodation in school, or understanding, while for others it may take the form of respected social boundaries, space to process difficult emotions, or acknowledgment of disability.

Social support has been found to be a factor that contributes to outcomes in individuals with CP and hEDS specifically (Kashikar-Zuck et al., 2008; Mu et al., 2019; Rombout et al.,

2010; Murray et al., 2013). As greater perceived social support has been associated with improved mental health outcomes in general pediatric populations (Chu et al., 2010) it is crucial to consider its relationship with the psychological wellbeing of those with pediatric hEDS.

Children with hEDS and their parents frequently report that social support is something that can make living with the syndrome easier (Bieniak, et al., Revise & Resubmit). However, a lack of social support has also been noted as being one of the hardest things about living with hEDS (Bieniak, et al., Revise & Resubmit). In addition, children with CP conditions report more struggles socially than their peers (Kashikar-Zuck et al., 2008; Mu et al., 2019). Similarly, in a study of adults with hEDS, only 52% of individuals felt supported by friends and 62% felt supported by their family (Murray et al., 2013). Adults with hEDS face more difficulties related to their health-related quality of life than peers without a CP condition, including perceptions of lower quality social support (Rombaut et al., 2010). The lack of social support for adults with the condition suggests that the social challenges experienced by those with pediatric hEDS are not simply a product of typical challenges associated with social development during childhood and adolescence but rather a long-term impact of hEDS.

The Relationship of Functional Disability, Social Support, and Mental Health

Functional Disability and Mental Health

At present, there is a gap of knowledge in understanding the impact of functional disability on mental health outcomes in individuals with pediatric hEDS. While it has been established that individuals with the syndrome do tend to have worse mental health outcomes (Cederlöf et al., 2016; Murray et al., 2013; Mu et al., 2019), it is unclear whether this is the consequence of functional disability alone or of a combination of other factors. Increased functional disability in similar CP conditions has been found to be related to worse mental health

outcomes (Simons et al., 2012), suggesting that a similar trend may be observed in individuals with pediatric hEDS. Furthermore, the more severe and frequent a child's pain is, the lower the quality of life is for that child in terms of general functioning psychological wellness (Hunfeld et al., 2001). When a child is experiencing more severe physical symptoms and increased functional disability, poorer mental health outcomes are to be expected. This highlights the necessity to find ways to manage the symptoms and associated disability in the hEDS population so that clinicians are best able create management programs and implement interventions that will yield the best possible outcomes for these individuals.

There is a positive association between psychological distress and functional disability in pediatric CP (Simons et al., 2012). As has been previously explained, those with hEDS experience chronic functional impairments, and these impairments may contribute to psychological struggles. Additionally, higher reports of anxiety and depression in childhood chronic pain are associated with worse pain outcomes (Pavlova et al., 2017), which may, in turn, create a cycle where the negative impacts of functional disability contribute to continued functional challenges. Fear of pain or injury, for example, is associated with increased functional disability, even at low pain levels, resulting in a need to discontinue engagement in previously pleasurable activities (Simons et al., 2012). Withdrawing from certain high-risk activities, such as contact sports, may be the best decision for reducing physical symptoms, however, loss of freedoms and ability to engage in activities that bring joy places an individual at risk for feelings of anger, sadness, and frustration (von Korff & Simon, 1996), all of which can contribute to negative psychological responses.

Functional Disability, Social Support, and Mental Health

Independently, the role of functional disability, social support, and mental health are each important aspects of pediatric hEDS. However, understanding these variables as being a part of a larger, complex system that influences the quality of life for those with the syndrome is necessary to be able to diagnose, treat, and cope with pediatric hEDS. Functional disability and social support have been found to not only be impacted by symptoms of CP and hEDS, but also to impact psychological outcomes themselves.

While there may be a direct influence of functional disability on mental health in hEDS, the relationship is undoubtedly influenced by additional factors. Social support is proposed in this study as being one of these factors, acting as a moderator in the relationship between functional disability and mental health. Social challenges are associated with worse mental health outcomes (Baeza-Velasco, et al., 2018; Rubin et al., 2001; López-Martínez, et al., 2008). This brings into question the influence of social support in the relationship between functional disability and mental health. As individuals with CP and hEDS tend to report lower levels of perceived social support than healthy peers this raises concerns for their mental health outcomes (Kashikar-Zuck et al., 2008; Mu et al., 2019). Adults with chronic pain who report higher levels of social support tend to report fewer depressive symptoms than those with lower levels of social support (López-Martínez et al., 2008). Furthermore, the same study of adults demonstrated no direct association between perceived social support and functional disability, suggesting that the two variables, while both important in the quality of life of chronic pain patients, are not significantly associated with one another (López-Martínez et al., 2008). As higher levels of social support appear to be a protective factor against negative psychological outcomes for children generally (Chu et al., 2010) and as social support itself is not correlated with functional

disability (López-Martínez et al., 2008), the impact of functional disability on mental health may be moderated by social support.

For children and adolescents, the type of social support, and the benefit of the social support, can vary greatly. For adolescents, social support is utilized as a way to cope with stressful situations (Camara et al., 2017). Yet, adolescents also report that certain support systems can act in ways that cause them distress (Camara et al., 2017). For example, while parents are often a source of significant support, they may also cause distress by encouraging high academic or extracurricular achievement (Camara et al., 2017). Similarly, peer relationships may become distressing if an adolescent feels they do not fit in with a desired social group (Camara et al., 2017). As a result, the specific domains of social support (e.g., parents, friends, teachers, classmates) matter less than the general perception that one is supported by those near to them.

In developing our model, we looked to the *Buffering Hypothesis* presented by Cohen and Wills (1985) and the *Disability-Stress-Coping Model* presented by Wallander and colleagues (1992, 1989). The *Buffering Hypothesis* (Cohen & Wills, 1985) posits that social support acts as a buffer against the negative impacts of chronic stress on individual well-being. In applying this model to our study, functional disability would act as a chronic stressor and social support would buffer the negative effects of FD on mental health. However, it is unclear if functional disability should be characterized as a chronic stressor, or as something that yields stress but is not within itself a stressor. To better understand the stress associated with functional disability, we looked to the *Disability-Stress-Coping Model* (Wallander et al., 1992, 1989) which presents social-ecological factors as resistance factors that mitigate the impact of risk factors on the outcome of biopsychosocial adaptations.

The Wallander et al. (1992, 1989) model is similar to the Cohen and Will's (1985) model in that it posits some chronic risk factors as being associated with wellness outcomes, with resistance factors influencing the relationship between said variables. The two models differ however, with the Cohen and Will's (1985) model presenting a moderation while the Wallander et al.'s (1992, 1989) model presenting a mediation. Further, the *Disability-Stress-Coping Model* is more complex in nature, including different subgroups of variables within each stage (i.e., risk factors include the subgroups of disease/disability parameters, functional care strain, and psychosocial stressors). Adding to the more complex nature of this model, the researchers demonstrate the presence of additional relationships between these subgroups of variables, such as a relationship between disease/disability parameters and psychosocial stressors.

In blending the two frameworks, we come to a model in which the relationship between functional disability, a risk factor per the *Disability-Stress-Coping Model*, and mental health outcomes, a well-being outcome per the *Buffering Hypothesis* and adaptation per the *Disability-Stress-Coping Model*, is impacted by social support, a buffer (Figure 1). As social support has been demonstrated to have a beneficial impact on mental health (Chu et al., 2010), but is not associated with functional disability (López-Martínez et al., 2008), we elected to follow the *Buffering Hypothesis*'s moderation model rather than the mediation presented in the *Disability-Stress-Coping Model*. Further, given the proposed moderation, we elect to utilize the language of “buffering”, presented by Cohen and Wills (1985), as well as “protective factors”, per Ann Masten (2003), as this language best characterizes both the main effect and interaction effect, we predict social support to have. Thus, it is proposed that social support will have a buffering effect such that increased social support will protect an individual with hEDS from some of the negative impacts of functional disability on mental health.

While the individual impacts of functional impairment vary between individuals, the need to alter one's behaviors as a consequence of functional impairment puts individuals into a situation where psychological distress can occur. Poor psychological outcomes can present additional challenges to individuals who are already may be struggling to cope with hEDS. Thus, the relationship between functional disability and mental health in hEDS has important influences on the quality of life of individuals with the condition. Further, the potential buffering effect of perceived social support against negative outcomes may present a mechanism by which, despite functional disability, individuals with pediatric hEDS experience better mental health outcomes than those lacking social support.

hEDS and Gender

It is important to understand if gender differences are observed in pediatric hEDS experiences. Previous research on hEDS has demonstrated that more females than males are diagnosed with the hEDS (Cederlöf et al., 2016) and that CP appears to be experienced differently by males and females (Keogh & Eccleston, 2006). Specifically, females report more pain (King et al., 2011; Keogh & Eccleston, 2006) and engage in more social support and internalizing behaviors while males engage more in distraction behaviors (Keogh & Eccleston, 2006). At present, there is no information available as to how gender may be related to mental health outcomes for individuals with hEDS. Yet, the presence of psychological struggles, as well as additional variables that put one at a higher risk for mental health challenges, it is important to evaluate additional factors that may increase susceptibility to psychological disorders. In exploring this relationship, we will begin to understand if different genders will experience hEDS in different ways, allowing future work to understand how, if differences do exist, gender influences the psychological outcomes of individuals with the condition.

The Present Study

The current study aims to improve our understanding of the relationships between functional disability, perceived social support, and mental health (anxiety and depression) in children with pediatric hEDS. The relationships between these variables will be assessed by the adaptation of the *Buffering Hypothesis* presented by Cohen and Wills (1985) and the *Disability-Stress-Coping Model* presented by Wallander et al. (1992, 1989). Our model proposes that the relationship between functional disability and mental health will be moderated by social support (Figure 1).

Hypothesis I is that increased functional disability will be associated with worse psychological outcomes, as measured by anxiety and depression symptoms. Hypothesis II is that social support acts as a moderator on this relationship. We will test the model above that proposes that increased levels of perceived social support will buffer the intensity of the impact of functional disability on anxiety and depression. Finally, exploratory analyses will build off of Hypothesis II, examining the role of gender in the relationships between variables of interest. Hypothesis III is that the correlations between functional disability and mental health will be significant for females and males, however those for females will be stronger given that females tend to utilize social support as a coping mechanism more frequently (Keogh & Eccleston, 2006).

Materials and Methods

Participants

The present sample includes 34 children and adolescents diagnosed with hEDS according to the Villefranche criteria (Beighton et al., 1998). These individuals were all invited to participate in the study while at a Chicago area EDS clinic. Participants ranged from 8 to 18

years old ($m = 14.68$, $sd = 2.9$). Six participants were children between 8 and 12 years old (17.65%), 10 were early adolescents between 13 and 15 years old (29.41%), and 18 were late adolescents between 16 and 18 years old (52.94%). Most participants identified as female ($n = 24$, 70.6%), with the remainder of the sample identifying as male ($n = 8$, 23.5%) and two people not reporting their gender. Twenty-eight participants provided information regarding their race and ethnicity, demonstrating that the sample was largely non-Hispanic white ($n = 18$, 52.94%). Additional racial and ethnic identities represented included white with Hispanic ethnicity ($n = 5$, 14.71%) as well as biracial identities of Asian or Asian American and white ($n = 2$, 5.88%), and American Indian or Alaskan Native and white ($n = 1$, 2.94%). Two individuals reported their ethnicity as Hispanic but did not report any racial identity. On average, it had been 3.5 years since children had attained their hEDS diagnosis ($sd = 2.81$). The means, standard deviations, and ranges for additional study variables are reported in Table 1.

Procedure

The study was completed with approval from the DePaul University Institutional Review Board (IRB) and the IRB of Advocate Children's Hospital. Following an appointment at a Chicago area EDS clinic, families were provided the opportunity to participate in the study. The study geneticist confirmed pediatric hEDS diagnosis prior to participation. Those families who volunteered to participate provided informed consent from the caregiver(s) and assent was given from their children. Caregivers and children each completed their respective forms at the clinic or returned the forms at a later date, with the option to mail back responses with pre-paid postage.

Materials

The present study is a part of a larger analysis of life with pediatric hEDS. At present we will analyze child reported functional disability, social support, mental health, pain, and gender.

Demographic Information. Information was gathered from both the child and the caregiver about the child's age, gender, race, and ethnicity.

Functional Disability. Functional disability was measured using the Functional Disability Inventory (FDI; Appendix A). This inventory was developed specifically for use in children and adolescents with chronic pain. The FDI consists of 15 questions, each rated on a 5-point Likert scale (0= "No trouble" and 4= "Impossible"), that measure the physical difficulty of day-to-day activities (Walker, & Greene, 1991). Questions cover these activities across different domains: home, school, recreation, and social. Examples of these include "Doing chores at home", "Being at school all day", "Doing activities in gym class (or playing sports)", and "Doing something with a friend (for example, playing a game)". The clinical utility of the measure has been validated for use in children with chronic pain conditions (Kashikar-Zuck, et al., 2011) and its reliability in our sample was excellent ($\alpha = .911$).

Mental health. Mental health was measured using the Pediatric Reported Outcomes Measurement Information System (PROMIS) Pediatric Anxiety Symptoms (Appendix B) and Depressive Symptoms (Appendix C) short forms. Each of the forms have been validated for pediatric CP populations as being accurate and precise measures of anxiety and depression symptoms (Irwin et al., 2010). PROMIS pediatric forms are developed for the use of individuals from age 8 to 18 (Varni et al., 2014). Each short form included 8 questions on a 5-point Likert scale (0= "Never", 4= "Almost always") to rate the child's frequency of experiencing symptoms of anxiety and depression over the last week. On both forms, total response scores ranged from 0

(not having experienced any of the symptoms over the past week) to 32 (having frequently experienced all of the symptoms over the past week). Questions on the PROMIS Anxiety Short Form include “I felt like something awful might happen” and “I felt worried”. Questions on the PROMIS Depression Short For include “I felt worthless” and “I felt like a failure”. Total scores for each form were converted to T scores for analysis, where the T score mean represents a general population of children ($M=50$, $SD=10$). The reliability scores in our sample were excellent for both the anxiety ($\alpha = .941$) and depression ($\alpha = .973$) questionnaires.

Social support. Social support was measured using the People in My Life (PIML; Appendix D) questionnaire. The PIML characterizes social support as how much an individual perceives that others care about their feelings and experiences, listen to and understand them, and treat them like a person (Ridenour et al., 2006; Harter, 2012). Social support, on this questionnaire, exists within four domains: parents, friends, classmates, and teachers. Examples of the questions on the form include choosing between the two phrases “Some kids have parents who don’t really understand them” and “Other kids have parents who really do understand them”. Then, for whichever phrase more closely matches their experience, the child marks the phrase as being “Really true for me” or “Sort of true for me”. Total overall scores are created by reverse scoring the appropriate questions, then taking the sum of all scores. Domain scores are created by taking the sum of all of the questions associated with the given domain. There were 24 questions in total on the PIML, and six in each domain, with possible overall scores ranging from 24 to 96. For the present study, we focused only on the measure of overall perceived social support. The reliability in our sample for the overall perceived social support was excellent ($\alpha = .909$). This measure has been found to be valid and reliable in the assessment of social support for individuals in late childhood, ages 10 to 12 years (Ridenour et al., 2006).

Pain. The intensity of pain was assessed by asking children “On days that you have had pain, what has been your usual level of pain in the last 2 weeks?”. Responses were provided on an 11-point numeric rating scale, with higher values representing more intense pain (0 = “*No Pain at All*”, 10 = “*Worst Pain I Can Imagine*”). The use of this question has been found to be a valid and reliable measure in pediatric populations (Castarlenas et al., 2017).

Gender. Child gender was collected via demographic questionnaire. Options provided for gender identification included female, male, and other.

Data Analysis

Descriptive and Demographic Analysis

Descriptive statistics will be conducted to examine the range, mean, and SD, of all variables in the model (functional disability, social support, and psychological outcomes). Additional descriptive statistics will be conducted to examine the range, mean, and SD of the age of participants. The percentages for race and sex will also be reported. SPSS will be utilized for data analysis except for Fisher’s Z which will be conducted with a publicly available, online software (Lowry, 2021).

Additionally, as CP is a common symptom of hEDS (Malfait et al., 2017), this study would not be complete without an analysis of the role of pain in child outcomes. Given that pain impacts all areas of an individual’s life, it is analyzed to best understand its potential role in the present model. Correlations between pain and each of the variables in the model (functional disability, social support, and mental health) will be conducted and if appropriate, pain may be considered a covariate in the subsequent analyses.

Hypothesis I Analysis

In order to examine the first hypothesis, which is that increased functional disability will be associated with worse psychological outcomes, Pearson correlation was utilized. Functional disability was measured using the total score on the FDI, and psychological outcomes (anxiety and depression) was measured using the T scores on the PROMIS anxiety and depression short forms. It was hypothesized that the correlations between functional disability and anxiety, and functional disability and depression would each be significant.

Hypothesis II Analysis

In order to examine the second hypothesis, which was that increased levels of perceived social support will mitigate the intensity of the impact of functional disability on psychological outcomes, a moderated regression analysis was conducted. The independent variable of functional disability was measured with the FDI, the dependent variable of mental health outcome was measured by the PROMIS anxiety and depression short forms, and the moderator of social support was measured with the PIML. As with the analysis for Hypothesis I, two different moderated regressions were run, the first looking at depression as an outcome and the second looking at anxiety.

Two moderated regressions were run separately to determine how social support moderates the relationships between functional disability and anxiety and depression, respectively. For each of the regressions, the FDI and PIML variables were centered. These centered scores were entered in the first step of the regression. The interaction between FDI and PIML was entered in the second step of the model.

To determine the appropriateness of conducting a moderated regression analysis to analyze how social support moderates the relationship between functional disability and mental

health, a power analysis was conducted to determine the minimum sample size needed. Power analysis determined that at 80% power, with a moderate to large effect size ($R^2 = 0.3$), a minimum sample of 30 participants will be needed. As the present sample meets this threshold ($n = 34$), a moderated regression analysis will be utilized. Additionally, past research has found a moderate to strong relationship between functional disability and mental health (Hershenfeld et al., 2016; Bulbena et al., 2012).

Hypothesis III: Gender

In terms of the number of males and females in the study, there were 24 females (70.6%) and 8 males (23.5%). This matches previous findings that females are more likely to be diagnosed with hEDS (Castori et al., 2010). What remains in question is if gender differences exist in childhood outcomes and relationships between variables. To analyze this, variable means were compared between genders using Welch's t-test for unequal samples. To analyze the differences in relationships between variables, correlations between FDI, PIML, anxiety, and depression were conducted for males and females independently. Then, the strength of relationships for each gender were compared using Fisher's z' Transformation.

Results

Correlations

As hypothesized, functional disability was found to have a significant, moderately strong, positive association with both mental health outcome variables (Anxiety $r = 0.53$, $p = 0.001$; Depression $r = 0.56$, $p = 0.001$). Functional disability and social support were not significantly correlated with one another ($r = -0.304$, $p = 0.109$). The association between depression and anxiety was significant, moderately strong, and positive ($r = 0.79$, $p < 0.001$). Social support was found to have a significant, moderately strong, negative association with anxiety ($r = -0.587$, $p =$

0.001) and a significant, moderately strong, negative association with depression ($r = -0.782$, $p = 0.0001$). This result would suggest that as social support increases, both anxiety and depression symptoms decrease.

Correlations Between Model Variables and Pain

Pain had a significant relationship with the predictor variable of functional disability and both mental health outcomes. The relationship between pain and functional disability was moderately strong and positive ($r = 0.68$, $p < 0.001$). The relationship between pain and anxiety was moderately strong and positive ($r = 0.50$, $p = 0.003$) and the relationship between pain and depression was moderately strong and positive ($r = 0.48$, $p = 0.004$). There was no significant relationship between pain and social support ($r = -0.267$, $p = 0.14$).

Given the relationships between pain and functional disability, pain and anxiety, and pain and depression, a hierarchical regression was run to determine if the proposed model would be better prepared to predict mental health outcomes if pain were included as a covariate.

Hierarchical regressions were run for both anxiety and depression, with pain as the first step (model 1) and functional disability as the second step (model 2). For both mental health outcomes, model 2, while significant models (anxiety $p = 0.002$; depression $p = 0.002$), were not significant improvements from model 1. Further analysis of model 2 demonstrated that neither pain nor functional disability were significant within the model for predicting anxiety or depression outcomes. These results suggest that while pain and functional disability are both significantly related to mental health, it does not appear that one variable is better suited to predict outcomes. Given these findings as well as past literature that demonstrates that pain and functional disability are closely related (Voermans et al., 2010; Simons et al., 2012), the original

study model, wherein the relationship between functional disability and mental health outcomes are moderated by social support, is retained.

Moderated Regression

The moderated regressions where social support was hypothesized to mitigate the impact of functional disability on mental health were found to be non-significant (See Table 2 for Depression and Table 3 for Anxiety). When anxiety was the outcome variable, functional disability and social support independently were significantly associated with anxiety (functional disability $p = 0.0001$; social support $p = 0.0016$), but the interaction between the two was not significant ($p = 0.3589$). When depression was the outcome variable, functional disability and social support independently were significantly associated with depression (functional disability $p = 0.00001$; social support $p = 0.00001$), but the interaction between the two was not significant ($p = 0.4335$).

Gender Differences

Exploratory analyses into the role of gender in mental health outcomes for individuals with pediatric hEDS were conducted via Welch's t-test, Pearson Correlations, and Fisher's Z.

First, Welch's t-test were conducted, given unequal sample sizes, to analyze differences between means for each variable between males and females. There was a significant gender difference for pain ($t = 7.127$, $p = 0.022$). No other variables differed significantly between gender (Table 4).

Second, Pearson Correlations between functional disability and mental health outcomes were conducted for males and females independently (Table 5). For males in the study ($n = 8$), the only significant correlation was between functional disability and depression, which was very strong and positive ($r = 0.967$, $p = 0.0001$). For females who provided complete data on both the

functional disability inventory and the PROMIS measures ($n = 22$), the only significant correlation was between function disability and anxiety, which was moderately strong and positive ($r = 0.482, p = 0.023$).

Pearson Correlations between social support and mental health outcomes were conducted for males and females. For males in the study, no significant correlations were detected. Further, for females who provided complete data on the social support measure and the PROMIS measures ($n = 21$), there was a significant, moderately strong, negative correlation between social support and anxiety ($r = -0.705, p = 0.0001$) and a significant, strong, negative correlation between social support and depression ($r = -0.897, p = 0.0001$).

Third, comparison of the correlations between genders utilizing Fisher's Z (Table 5) demonstrated that the genders differed significantly on their correlations between functional disability and depression ($Z = 3.33, p = 0.0009$) and social support and depression ($Z = 2.109, p = 0.0349$). The relationship between functional disability and depression was significant for males but not for females while the relationship between social support and depression was significant for females but not for males. No significant differences were seen between functional disability and anxiety or social support and anxiety.

Discussion

In order to better understand the role of social support in living with pediatric hypermobile Ehlers-Danlos Syndrome, the present study analyzed the role of social support in the relationship between functional disability and mental health in individuals with pediatric hEDS. The Cohen and Wills *Buffering Hypothesis* (1985) and the *Disability-Stress-Coping Model* presented by Wallander et al. (1992, 1989) were utilized to create a new model for considering the relationships between the present variables. In combining these models, we

developed our own in which functional disability acts as risk factor (Wallander et al., 1992, 1989), social support acts as a buffer/moderator (Cohen & Wills., 1985; Wallander et al., 1992, 1989), and mental health, as measured by anxiety and depression symptoms, is a wellness outcome/adaptation (Cohen & Wills., 1985; Wallander et al., 1992, 1989).

Support was found for Hypothesis I, which proposed that as functional disability increased, so would symptoms of anxiety and depression. However, support was not found for Hypothesis II, which suggested that social support acts as a moderator on the relationship between functional disability and mental health. These findings contradict the prediction that increased levels of social support would act as a buffer against the negative impacts of functional disability on mental health. Finally, mixed results were found for Hypothesis III, which proposed that while significant correlations would be found for both males and females between functional disability and mental health variables, the correlations would be stronger for females given their tendency to utilize social support more often as a coping mechanism (Keogh & Eccleston, 2006).

Hypothesis I: Relationships between Functional Disability, Social Support, and Mental Health

Within the present sample, as functional disability increased so did symptoms of both anxiety and depression. Such findings are consistent with past research in general pediatric CP samples that demonstrate an association between functional disability and general mental health outcomes (Vinall et al., 2016; Pavlova et al., 2017). More specifically, the relationship between functional disability and anxiety has been documented in pediatric CP populations (Simons et al., 2012) and the relationship between functional disability and depression has been documented in general CP populations (Fishbain et al., 1997). As high levels of functional disability are frequent in hEDS (Voermans et al., 2010; Syx et al., 2017), these associations are concerning for

the psychological wellness of those with the syndrome. These findings would suggest that clinicians should engage in ongoing assessment of functional disability and mental health in individuals with pediatric hEDS. Further, if an individual is reporting high levels of functional disability or mental health concerns, it would be in the best interest of the child to explore the other variable as well (e.g., if a child is reporting that they are struggling to engage in daily tasks, the clinician should also assess their mental health).

Functional disability was not significantly associated with social support. These results are similar to those in adults, where functional disability and social support are not significantly associated with one another (López-Martínez et al., 2008). Social support was significantly associated with anxiety and depression. This result reinforces previous findings that have demonstrated that increased social support is related to better mental health outcomes in children (Chu et al., 2010) and that lower social support is related to worse mental health outcomes generally (Baeza-Velasco, et al., 2018; Rubin et al., 2001; López-Martínez, et al., 2008).

Hypothesis II: The Moderation Model

Hypothesis two was not supported by the results of the moderated regression analysis. The results demonstrate that functional disability and social support are both independently related with mental health outcomes, but social support does not moderate the relationship between functional disability and those outcomes. These results suggest that regardless of social support, functional disability is significantly associated with mental health outcomes, such that increased functional disability is related to more frequent symptoms of anxiety and depression.

There are a few reasons why our hypothesis may have not been supported. First, we utilized our measure to observe overall social support, including that from teachers, classmates, friends, and parents (Ridenour et al., 2006; Harter, 2012). Given that general social support had

been previously demonstrated to be related to mental health outcomes (Baeza-Velasco, et al., 2018; Rubin et al., 2001; López-Martínez, et al., 2008), we felt that overall social support may also have been influential in the relationship between functional disability and mental health. However, it may be that specific sources of social support, such as friends or parents, maybe more substantial in their support of the child, while others, such as classmates or teachers, may be more limited, resulting in essentially a balancing out of these levels in the child's overall rating of their social support. Unfortunately, our present sample was not large enough to examine potential differences of source of social support.

Second, although social support generally has a buffering effect, there are situations in which social support poses a threat to well-being. Miscarried helping is one example of this phenomenon. Miscarried helping is observed when a caregiver of a child with a disability attempts to assist their child in their coping, however this helping results in inadvertently harming the child's self-efficacy or in frustration with the parent's actions, thus damaging the efficacy of the parental support (Fales et al., 2014; Harris et al., 2008). For example, a parent of a child with hEDS may consistently try to help them with daily tasks like making food or doing their laundry. The parent may jump in to help the child as they do the task, in an attempt to ease the burden on the child. In contrast to the parent's goal, the child may feel frustrated that their parent feels they are unable to complete the task, eventually expecting the parent to step in to assist so they avoid the task altogether. In contrast, miscarried helping may also result in a child feeling that a task is something that they are unable to complete independently, resulting in increases in the level of the child's perceived functional disability or in their actual functional disability if they become psychologically conditioned to ask for help or physically decondition to

be able to do the task. Consequently, the parent's helping has caused frustration and/or increased the child's perceived functional disability.

The positive correlation between child perceptions of parent miscarried helping and child perception of poor family functioning demonstrates the negative influence of miscarried helping on parental support (Fales et al., 2014). Further, higher levels of disability are related to perceived miscarried helping (Fales et al., 2014), which, given the high levels of functional disability in individuals with pediatric hEDS (Voermans et al., 2010), is a concern for our sample. As such, miscarried helping may partially explain why social support was not observed as a moderator in the present study.

Hypothesis III: Exploring the Role of Gender in hEDS

The results of our gender exploratory analyses present differences between males and females, which furthers evidence that gender identity may influence CP experiences (Keogh & Eccleston, 2006). First, in contrast to our hypothesis, when considering the relationships between functional disability and mental health outcomes, only males demonstrated a significant, positive correlation between functional disability and depression. This relationship was not significant in females, and the difference between the correlations in males and females was significant. Second, when considering the relationships between social support and mental health outcomes, only females demonstrated significant, negative correlations between social support and both mental health variables; however, only the relationship between functional disability and depression was significantly stronger for females when compared to males.

These results fit within the current literature that claims that social support plays a larger role for females with CP than for their male counterparts (Keogh & Eccleston, 2006). Similar trends were observed in a recent meta-analysis by Chu and colleagues (2010), where they

determined that the impact of social support on general well-being in pediatric individuals was significantly larger for females than males. Additional research by Zhang and colleagues (2015) finds that the interaction between stress and friend support was significantly related to depression for females but not males. The findings of these previous studies demonstrate that our results, despite the small sample, are likely reflective of differences in the lived experience of hEDS across genders.

There were also significant gender differences between functional disability and depression. One potential explanation for this finding is that the masculine behavioral expectations for boys in Western culture includes being independent and having minimal reliance on support from others (Pollack, 2006). This “Boy Code” adds increasing pressure to young males as they age, resulting in feelings of loneliness, a pressure to express their true emotional experiences, and social expectations to act tough (Pollack, 2006). Applying this framework to the present study would suggest that when males with pediatric hEDS are experiencing increased functional disability they may also be experiencing the weight of not meeting societal expectations of toughness. It is possible that this pressure experienced by males to meet masculine expectations paired with the inability to do so at times because of functional disability yields feelings of isolation, hopelessness, and poor self-esteem – all of which would put someone at a higher risk for depressive symptoms. Additionally, the pressure to not engage in social support the way that females are expected to may explain in part why we see a significant relationship between social support and depression in females, but not males.

Furthering this research, other studies have determined that a persistent stereotype surrounding the experience of pain is that men, compared to females, tend to have more endurance in coping with pain (Wallander et al., 2012; Robinson et al., 2001). In both of these

studies, both males and females endorsed this stereotype (Wallander et al., 2012; Robinson et al., 2001). These widely held beliefs regarding the experiences of males and pain likely play a role in the significant difference between males in females in the relationship between functional disability and depression.

The variability in the relationships between functional disability, social support, and mental health, the lack of consensus on the influence of gender in hEDS, and the increased presence of mental health challenges in those with health struggles together highlight the need for ongoing exploration of gender differences in the experience of pediatric hEDS. As demonstrated in our findings, there appear to be differences in the experience of hEDS in relation to gender. These findings do not solve the ongoing debate regarding the role of gender in hEDS, but rather reaffirm the need to examine gender as we continue to study the impacts of and interventions for pediatric hEDS.

Clinician Implications

In considering the clinical implications of this study, we must look at both the associations between variables and the absence of moderation. Clinically, correlation analyses demonstrate a need to emphasize the importance of social support in children with hEDS for the sake of their psychosocial wellbeing. To facilitate social wellbeing, care providers can educate patients and families on the importance of social support and the different sources of support, work with families to ensure that they are best equipped to support the child and one another, and facilitate conversations with children about how to maintain healthy relationships.

Additionally, the correlation analyses demonstrate that a lower level of functional disability in children with hEDS may be associated with fewer mental health difficulties. Further, the perception of having sufficient, quality social support may also be related to fewer mental

health difficulties but is likely unrelated to the child's level of functional disability. These findings reassert the need for care providers to attend to not only the primary symptoms of hEDS, but also the additional impacts of the condition including functional disability, social support, and mental health.

While the management of functional disability and social support are important in relation to the mental health of individuals with pediatric hEDS, social support does not moderate the relationship between functional disability and anxiety or depression. From a clinical viewpoint, this highlights the need to attend to these variables as being independently related to the psychological wellbeing of a person with pediatric hEDS. Providing resources or interventions designed to improve social support, while related to fewer symptoms of anxiety and depression, will not mitigate the relationship between functional disability on mental health.

In sum, clinicians should attend to not only the primary symptoms of hEDS but the functional and psychosocial consequences of the condition as well. Further, in attending to these additional impacts, clinicians should recognize that successful social interventions will not correspond with decreased functional disability. However, improved social support is likely to coincide with improvement in symptoms of anxiety and depression.

Limitations and Future Directions

There are a few key limitations to be considered when thinking of the results of this study. First, as a consequence of the small sample size, these findings only represent the experiences of a small subset of individuals with hEDS. In particular, most of the participants came from middle to upper class families, had private insurance, and identified as White. Consequently, there is little understanding of the role of functional disability and social support in the lives of children with hEDS who have been marginalized or minoritized. As hEDS and CP

impact individuals across all backgrounds, future work must ensure that samples represent such diversity.

An additional limitation of the present study is in the assessment of the directionality of the relationships. As the FDI, PIML, and PROMIS measures all have overlapping time frames from which they assess symptoms, it is unclear as to the chronological direction of the onset of variables. Thus, it is unreasonable to assert that the direction of the relationships proposed here is the only potential mode of interaction between these variables. Further complicating these relationships, previous work with adolescents with CP has found that a portion of participants report having mental health disorders prior to the onset of their CP (Tegethoff et al., 2015). Thus, future studies should assess onset of not only CP symptoms, but the onset of psychological symptoms as well. The addition of such data would allow for differentiation between individuals who had challenges with their mental health prior to CP onset, those who have had challenges with their mental health following CP onset, and those who have not had significant mental health challenges prior to or following CP. Additionally, there may be some sort of feedback system that exists between variables. For example, anxiety surrounding pain or risk of injury could result in withdrawal from activities, increasing feelings of social isolation, while depression could result in feelings of hopelessness that make even the thought of beneficial treatments, such as physical therapy (Levy, 2018), feel pointless. Future studies should engage in more longitudinal work to allow for the assessment of the directionality of variables and their potential bidirectional or reciprocal relationships. Assessment of directionality and potential feedback systems will allow for a more thorough understanding of the consequences of the complex condition.

Given that social support can provide both assistance and challenges to youth (Camara et al., 2017; Bieniak et al., Revise & Resubmit) and that there are many domains from which it originates (Ridenour et al., 2006), future work should analyze the role of different sources of social support in the wellbeing of this population. It is possible that by looking at social support broadly in this study, we missed important influences of the specific kinds of support a child has in their life. While analysis of average social support is important, we encourage more specific analyses in the future.

Our exploratory analyses demonstrate the need for continued study of the role gender plays in the experience of CP and hEDS specifically. In particular, future studies should assess the relationship between study variables in a larger cohort of participants. Additionally, in larger studies a broader range of gender identities should also be assessed so that the experiences of individuals who identify as a gender other than male or female or who do not identify as a particular gender can be understood.

Conclusion

The present study demonstrates the complexity of life with pediatric hypermobile Ehlers-Danlos Syndrome. Results emphasize the need for illness management to be multidisciplinary, such that those with the condition are able to find ways to cope with the physical and psychosocial consequences of the syndrome. As the symptomology of hEDS varies between individuals, it is crucial to continue to investigate potential individual differences or factors as they relate to mental health outcomes. Early intervention that aims to minimize the functional disability, social challenges, and psychological distress of those with pediatric hEDS should relate to improved overall wellbeing of the child over their lifetime.

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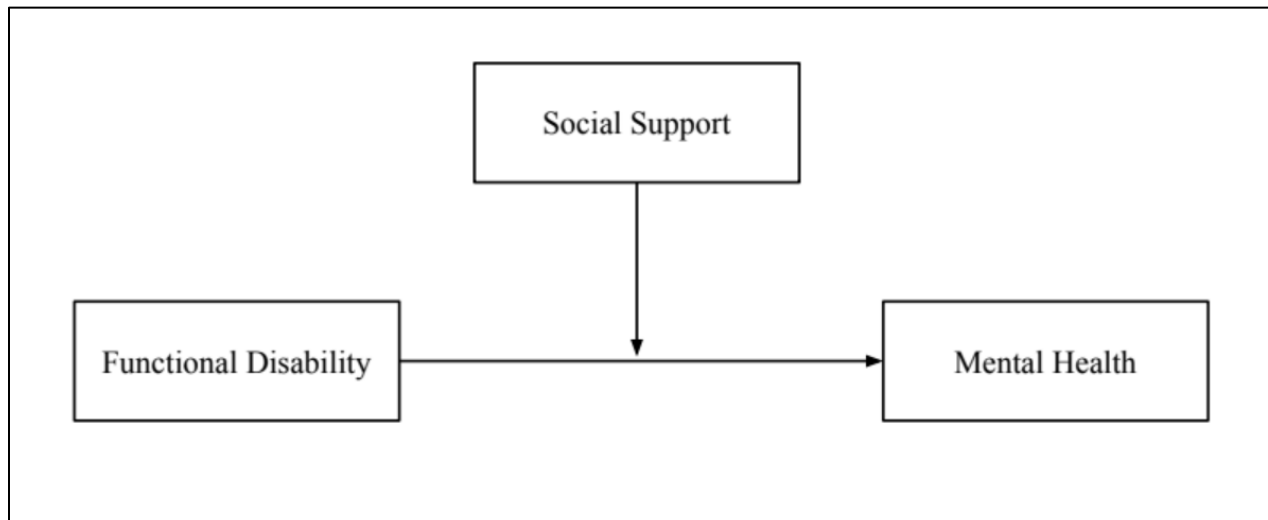
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Figure 1

Proposed Model



Note. The proposed model, based on the Cohen and Wills *Buffering Hypothesis* (1985) and the Wallander et al. (1992, 1989) *Disability-Stress-Coping Model*; Functional disability is the risk factor, mental health is the wellness outcome, and social support is the buffer that moderates the negative impact of functional disability on mental health.

Table 1*Descriptive Statistics for Study Variables*

Variable	Mean	Standard Deviation	Range	N
PROMIS Anxiety	56.585	13.3158	49.3 (33.5-83.3)	34
PROMIS Depression	54.138	14.3523	47.2 (35.2-82.4)	34
FDI	19.1563	11.4416	40 (0-40)	32
PIML	79.3667	11.6159	39 (56-95)	30
Pain	5.3235	6.6021	8 (0-8)	34

Table 2*Moderated Regression: Depression*

					95% Confidence interval	
	<i>B</i>	<i>SE</i>	<i>t</i>	<i>p</i>	Lower	Upper
Constant	54.25	1.27	42.67	<.00001**	51.63	56.87
FDI	0.57	0.12	4.89	<.00001**	0.33	0.81
PIML	-0.80	0.11	-7.23	<.00001**	-1.03	-0.57
Interaction (FDI x PIML)	-0.01	0.01	-0.80	.43	-0.03	0.01

Note. Significant statistics denoted by * for $p < 0.05$ and ** for $p < 0.01$.

Table 3*Moderated Regression: Anxiety*

					95% Confidence interval	
	<i>B</i>	<i>SE</i>	<i>t</i>	<i>p</i>	Lower	Upper
Constant	56.4223	1.7638	31.9885	<.00001**	52.7895	60.0551
FDI	0.5985	0.1621	3.6931	.0011**	0.2647	0.9323
PIML	-0.5426	0.1538	-3.5273	.0016 **	-0.8595	-0.2258
Interaction (FDI x PIML)	-0.0120	0.0129	-0.9348	.3589	-0.0385	0.0145

Note. Significant statistics denoted by * for $p < 0.05$ and ** for $p < 0.01$

Table 4*Descriptive Statistics by Gender*

Variable	Mean (<i>SD</i>)		Range		Welch's t- test between genders	<i>p</i>
	<i>Male</i>	<i>Female</i>	<i>Male</i>	<i>Female</i>		
PROMIS Anxiety	48.438 (11.9649)	58.687 (13.3288)	29.9 (33.5- 63.4)	45.3 (38- 83.3)	4.153	.062
PROMIS Depression	47.525 (13.7837)	56.363 (14.7415)	32 (35.2- 67.2)	47.2 (35.2- 82.4)	2.381	.147
FDI	12.75 (15.71851)	20.5455 (9.10116)	40 (0-40)	36 (0-36)	1.754	.219
PIML	83.12 (9.7165)	77.4762 (12.1886)	25 (70-95)	39 (56-95)	1.690	.212
Pain	3.1250 (2.64237)	5.916 (2.30154)	7 (0-7)	8 (0-8)	7.127	.022*

Note. Significant statistics denoted by * for $p < 0.05$. For males, N=8 for all variables. For females, N=24 for PROMIS Anxiety, PROMIS Depression, and Pain, N=22 for FDI, and N=21 for PIML.

Table 5*Pearson Correlations and Fisher's Z by Gender*

Variables correlated	<i>Male</i>		<i>Female</i>		Fisher's Z	
	<i>r</i>	<i>p</i>	<i>r</i>	<i>p</i>	<i>Z</i>	<i>p</i>
FDI - Anxiety	0.565	.144	.482	.023*	0.23	0.8181
FDI - Depression	.967	.0001**	.354	.106	3.33	0.0009**
FDI – PIML	-.273	.513	-.361	.118	0.194	0.8493
PIML - Anxiety	-.079	.852	-.705	<.0001**	1.579	0.11410
PIML - Depression	-.372	.364	-.897	<.0001**	2.109	0.0349*

Note. Significant statistics denoted by * for $p < 0.05$ and ** for $p < 0.01$. For males, N=8 for all variables. For females, N=22 for FDI-Anxiety and FDI- Depression, and N=21 FDI-PIML, PIML-Anxiety, and PIML-Depression.

Appendix A

Functional Disability Inventory

Child and Adolescent Form

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.	0	1	2	3	4
2. Walking up stairs.	0	1	2	3	4
3. Doing something with a friend. (For example, playing a game.)	0	1	2	3	4
4. Doing chores at home.	0	1	2	3	4
5. Eating regular meals.	0	1	2	3	4
6. Being up all day without a nap or rest.	0	1	2	3	4
7. Riding the school bus or traveling in the car.	0	1	2	3	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.	0	1	2	3	4
9. Doing the activities in gym class (or playing sports).	0	1	2	3	4
10. Reading or doing homework.	0	1	2	3	4
11. Watching TV.	0	1	2	3	4
12. Walking the length of a football field.	0	1	2	3	4
13. Running the length of a football field.	0	1	2	3	4
14. Going shopping.	0	1	2	3	4
15. Getting to sleep at night and staying asleep.	0	1	2	3	4

The Functional Disability Inventory can be accessed at

https://www.researchgate.net/publication/21330310_The_Functional_Disability_Inventory_Measuring_a_Neglected_Dimension_of_Child_Health_Status

Walker, L.S., & Greene, J. W. (1991). The functional disability inventory: Measuring a neglected dimension of child health status. *Journal of Pediatric Psychology, 16*(1), 39-58.

<https://doi.org/10.1093/jpepsy/16.1.39>

Appendix B

PROMIS® Pediatric Anxiety Short Form

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

Subject ID # _____

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.					

The PROMIS Pediatric Anxiety Short Form can be accessed at

<https://www.healthmeasures.net/search-view-measures?task=Search.search>

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Appendix C

PROMIS® Pediatric – Depression Short Form

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.					

The PROMIS Pediatric Depression Short Form can be accessed at

<https://www.healthmeasures.net/search-view-measures?task=Search.search>

Irwin, D. E., Stucky, B., Langer, M. M., Thissen, D., Dewitt, E. M., Lai, J. S., Varni, J. W.,

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Appendix D

People In My Life

Subject ID # _____

There are two steps to answering each question below:

- 1) In each numbered row, decide whether you are more like the kids on the left side or on the right side.
- 2) Then, for the side you picked, decide whether the statement is Sort of True for you or Really True for you.

Put an X in the box that best describes you.

There should only be 1 box checked per row. You may pick different sides for different questions.

	Really True for me	Sort of True for me			Sort of True for me	Really True for me	
Sample Sentence							
a.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids like to do fun things with a lot of other people	BUT	Other kids like to do fun things with just a few people	<input type="checkbox"/>	<input type="checkbox"/>
1.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids have parents who don't really understand them	BUT	Other kids have parents who really do understand them	<input type="checkbox"/>	<input type="checkbox"/>
2.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids have classmates who like them the way they are	BUT	Other kids have classmates who wish they were different	<input type="checkbox"/>	<input type="checkbox"/>
3.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids have a teacher who helps them if they are upset or have a problem	BUT	Other kids don't have a teacher who helps them if they are upset or have a problem	<input type="checkbox"/>	<input type="checkbox"/>
4.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids have a close friend who they can tell problems to	BUT	Other kids don't have a close friend who they can tell problems to	<input type="checkbox"/>	<input type="checkbox"/>
5.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids have parents who don't seem to want to hear about their children's problems	BUT	Other kids have parents who do want to listen to their children's problems	<input type="checkbox"/>	<input type="checkbox"/>
6.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids have classmates they can become friendly with	BUT	Other kids don't have classmates that they can become friendly with	<input type="checkbox"/>	<input type="checkbox"/>
7.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids don't have a teacher who helps them to do their very best	BUT	Other kids do have a teacher who helps them to do their very best	<input type="checkbox"/>	<input type="checkbox"/>
8.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids have a close friend who really understands them	BUT	Other kids don't have a close friend who really understands them	<input type="checkbox"/>	<input type="checkbox"/>
9.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids have parents who care about their feelings	BUT	Other kids have parents who don't seem to care very much about their feelings	<input type="checkbox"/>	<input type="checkbox"/>
10.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids have classmates who sometimes make fun of them	BUT	Other kids don't have classmates who make fun of them	<input type="checkbox"/>	<input type="checkbox"/>

	Really True for me	Sort of True for me			Sort of True for me	Really True for me	
11.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids do have a teacher who cares about them	BUT	Other kids don't have a teacher who cares about them	<input type="checkbox"/>	<input type="checkbox"/>
12.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids have a close friend who they can talk to about things that bother them	BUT	Other kids don't have a close friend who they can talk to about things that bother them	<input type="checkbox"/>	<input type="checkbox"/>
13.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids have parents who treat their child like a person who really matters	BUT	Other kids have parents who don't usually treat their child like a person who matters	<input type="checkbox"/>	<input type="checkbox"/>
14.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids have classmates who pay attention to what they say	BUT	Other kids have classmates who usually don't pay attention to what they say	<input type="checkbox"/>	<input type="checkbox"/>
15.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids don't have a teacher who is fair to them	BUT	Other kids do have a teacher who is fair to them	<input type="checkbox"/>	<input type="checkbox"/>
16.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids don't have a close friend who they like to spend time with	BUT	Other kids do have a close friend who they like to spend time with	<input type="checkbox"/>	<input type="checkbox"/>
17.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids have parents who like them the way they are	BUT	Other kids have parents who wish their children were different	<input type="checkbox"/>	<input type="checkbox"/>
18.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids don't get asked to play in games with classmates very often	BUT	Other kids often get asked to play in games by their classmates	<input type="checkbox"/>	<input type="checkbox"/>
19.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids don't have a teacher who cares if they feel bad	BUT	Other kids do have a teacher who cares if they feel bad	<input type="checkbox"/>	<input type="checkbox"/>
20.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids don't have a close friend who really listens to what they say	BUT	Other kids do have a close friend who really listens to what they say	<input type="checkbox"/>	<input type="checkbox"/>
21.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids have parents who don't act like what their children do is important	BUT	Other kids have parents who do act like what their children do is important	<input type="checkbox"/>	<input type="checkbox"/>
22.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids often spend their recess being alone	BUT	Other kids spend recess playing with their classmates	<input type="checkbox"/>	<input type="checkbox"/>
23.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids have a teacher who treats them like a person	BUT	Other kids don't have a teacher who treats them like a person	<input type="checkbox"/>	<input type="checkbox"/>
24.	<input type="checkbox"/>	<input type="checkbox"/>	Some kids don't have a close friend who cares about their feelings	BUT	Other kids do have a close friend who cares about their feelings	<input type="checkbox"/>	<input type="checkbox"/>

Susan Harter, Ph.D., University of Denver, 2012

The People in My Life questionnaire can be accessed at

https://d1wqtxts1xzle7.cloudfront.net/52927378/Social_Support_for_Children-with-cover-page.pdf?Expires=1621970983&Signature=gOynKIL-

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