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# Upper Extremity Kinematics and Pain Outcomes During Activities of Daily Living in Pediatric Hypermobile Ehlers-Danlos Syndrome

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# UPPER EXTREMITY KINEMATICS AND PAIN OUTCOMES DURING ACTIVITIES OF DAILY LIVING IN PEDIATRIC HYPERMOBILE EHLERS-DANLOS SYNDROME

by

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# ABSTRACT

UPPER EXTREMITY KINEMATICS AND PAIN OUTCOMES DURING ACTIVITIES OF DAILY LIVING IN CHILDREN WITH HYPERMOBILE EHLERS DANLOS SYNDROME
by

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Under the Supervision of Dr. Brooke A. Slavens, PhD

Ehlers-Danlos syndrome (EDS) is a group of heritable connective tissue disorders, consisting of thirteen different subtypes. Among the thirteen, Hypermobile Ehlers Danlos Syndrome (hEDS) is the most common. Individuals with this condition present with frequent joint instability that results in ongoing subluxations and dislocations. Secondary diagnoses of this condition include chronic pain, Myalgic Encephalomyelitis/Chronic Fatigue Syndrome (ME/CFS), Major Depressive Disorder (MDD), and Generalized Anxiety Disorder (GAD). The diagnostic process of hEDS is convoluted by the lack of a genetic identifier (Malfait et al., 2017). Individuals with hEDS are often not diagnosed until later in adulthood, leaving their adolescent life full of confusion and difficulty in managing the symptoms. As a result, children with hEDS endure a childhood of difficulty in participating in school, activities, and engaging with peers The primary aims of this study were to characterize upper extremity kinematics during activities of daily living, to characterize pain location and severity, to understand common treatments for pain management, and to understand how pain severity can impact the child's self-perception of disability due to pain in children with hEDS. This study included 11 children with hEDS. Kinematic data collection consisted of using a retroreflective marker set and 3D motion capture system. To obtain data on pain, a non-standardized Pain Severity and Location Questionnaire

and Numerical Rating Scale (NRS) (Breivik et al., 2008) were administered interview style. Obtaining data on treatments, a non-standardized Treatment History Questionnaire was administered to the child's legal guardian. The Functional Disability Inventory (FDI) was administered to identify the child's level of perceived disability due to his or her pain (Kashikar-Zuck et al., 2011) Obtaining kinematic data of a normative pediatric population was restricted due to Coronavirus Disease (COVID-19). Existing literature was utilized to identify clinical differences, however, due to the variability between research this study cannot conclude whether children with hEDS have a greater range of motion than a normal pediatric group. However, this study identified that the four ADL tasks can be completed with wrist flexion and extension within values of 12° and 56° respectively and wrist ulnar and redial deviation within values of 20° and 11° respectively. At the shoulder, the tasks can be completed within 55° of shoulder flexion and 46° of extension. Within the four tasks, internal rotation required maximal of 42° and external rotation required 94°. At the thoracohumeral joint, maximal shoulder flexion was 142° and abduction was 57° during the combing task. Internal rotation was greatest during the reaching across task, reaching 81°. Data on pain location and severity displayed results that all children in this study reported pain in more than one bodily location. Further, all of the children in this study reported pain in their back. When describing their worst location of pain, all of the children in this study reported pain levels greater than or equal to 4/10, designating moderate to severe pain throughout the entire group. The maximal amount of treatments trialed was 10 while the least amount was 0. Parents reported a mode answer of 'satisfied' when questioned their level of satisfaction with treatments trialed for pain on a Likert satisfaction scale. The scores from the Functional Disability Inventory ranged from minimal to severe perceived disability. However, higher FDI scores did not correlate with higher NRS scores. movement patterns has the potential

to assist in identifying phenotypic characteristics of this group, which in turn could inform treatment practices and guide future research in identifying a genotype. Additionally, obtaining information regarding chronic pain, fatigue, and psychological experiences in pediatric hEDS can inform researchers on pediatric specific symptoms to more accurately diagnose this population.

I would like to dedicate this thesis to all the children that participated in this study, your vigor and resiliency is incomparable. I would like to offer a special dedication to my parents as they have always instilled the value of education and research in our home.

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| | |
|---|---|
| EDS | Ehlers-Danlos Syndrome |
| hEDS | Hypermobile Ehlers-Danlos Syndrome |
| HCTD | Heritable Connective Tissue Disorder |
| ADL | Activities of Daily Living |
| AOTA | American Occupational Therapy Association |
| GH | Glenohumeral |
| TH | Thoracohumeral |
| OT | Occupational Therapy |
| UE | Upper Extremity |
| ROM | Range of Motion |
| GAD | Generalized Anxiety Disorder |
| MDD | Major Depressive Disorder |
| ME/CFS | Myalgic Encephalomyelitis/Chronic Fatigue Syndrome |
| TFCC | Triangular Fibrocartilage Complex |
| FDI | Functional Disability Inventory |
| NRS | Numerical Rating Scale |

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# I. INTRODUCTION

Ehlers-Danlos Syndrome (EDS) consists of a group of heritable connective tissue disorders, comprised of 13 different subtypes. Each subtype of EDS presents with joint hypermobility, skin hyperextensibility, and tissue fragility. However, each subtype will vary with which bodily system the condition manifests and has a unique identified genotype for diagnostic certainty. Hypermobile Ehlers-Danlos Syndrome (hEDS) is the only subtype without an identified genotype, resulting in a greater difficulty for diagnosis and effective treatment (Malfait et al., 2017).

At present, it is estimated that E.DS occurs 1/5,000 people, with hEDS comprising 80% of all EDS cases (Tinkle et al., 2017). Individuals with hEDS present with frequent joint subluxation and dislocations, chronic pain, fatigue, depression, and anxiety (Gurley-Green, 2001; Malfait et al., 2017). Due to the lack of an identified genotype, hEDS is often mistaken for other medical conditions or ignored throughout childhood, leading to further health complications and more intense symptoms in adulthood (Gurley-Green, 2001). Current research has shown that children with hEDS have difficulty in participating in their instrumental and basic activities of daily living (ADL) (AOTA, 2020). More specifically, the hEDS pediatric population has displayed problems in performing in school, participating in extracurricular activities, and engaging with their peers (Engelbert et al., 2017).

Characterizing glenohumeral joint kinematics during ADL movement patterns has the potential to assist in identifying phenotypic characteristics of this group, which in turn could inform treatment practices and guide future research in identifying a genotype. Additionally, obtaining information regarding chronic pain, fatigue, and psychological experiences in pediatric hEDS can inform researchers on pediatric specific symptoms to more accurately diagnose this

population. The primary goals of this research study are to characterize pediatric glenohumeral (GH) kinematics and identify whether pain is a primary symptom in the pediatric population of hEDS.

**Statement of the Problem**

Hypermobile Ehlers-Danlos Syndrome (hEDS) is the only subtype of the Ehlers Danlos syndromes without an identified genotype, causing heightened difficulties for providing sound diagnoses (Beighton, De Paepe, Steinmann, Tsipouras, & Wenstrup, 1997; Malfait et al., 2017). At present, the majority if hEDS patients are unaware they have this condition until adulthood, leaving their adolescent years filled with the strain of enduring the symptoms independently and without effective medical intervention (Tinkle et al., 2017). Current research surrounding hEDS should focus on identifying a specific genotype, evaluating the most effective treatment options for hEDS patients, and compiling phenotypical kinematic traits. Adults with hEDS frequently experience chronic pain and fatigue, anxiety, and depression in addition to their musculoskeletal symptoms (Tinkle et al., 2017). There is less research available on the pediatric presentation, symptoms, and lived experience of hEDS (Engelbert et al., 2017). However, it is known that hEDS is multifaceted and often negatively impacts the individual's ability to fully participate in their daily life. Researching the specific GH kinematics in children with hEDS in relation to their pain outcomes may surface previously unknown correlations that could guide future treatment of hEDS. Results from this study can further guide practice for children and adults with hEDS so that they are provided more accurate diagnosis and effective treatment.

**Purpose**

The primary purposes of this research study are to characterize GH joint kinematics during activities of daily living (ADL) of children diagnosed with hEDS, to analyze the presence

and severity of pain, to identify the level of perceived disability due to pain, and to find common treatment modalities and level of satisfaction with treatment. The GH joint is used often, as it offers the widest range of motion in the upper extremity. Identifying and characterizing the joint kinematics at the GH joint could provide insight into phenotypical presentation in pediatric hEDS and can identify why. Additionally, understanding the severity of pain in this pediatric population may allow clinicians to recognize pain characteristics and to have a better understanding in how to treat the pain.

**Hypothesis and Aims**

The primary objective within this research study is to characterize the glenohumeral joint kinematics in children with Hypermobile Ehlers-Danlos syndrome and to obtain a clear understanding of their pain severity, interference, and management. The aims listed below support the primary objectives of this research.

*Aim 1*: To characterize upper extremity 3D kinematics during activities of daily living in pediatric hEDS. This study will investigate four movement components of ADL including combing, drinking, reaching across the midline, and reaching back to the ipsilateral side. Glenohumeral, thoracohumeral, and wrist kinematics will be measured utilizing Vicon Motion Capture system (Oxford Metrics) with a set of 27 reflective markers. It is hypothesized that during the ADL, children with hEDS will display greater glenohumeral, thoracohumeral, and wrist range of motion (ROM) when compared to existing kinematic literature. Research has identified greater active range of motion in glenohumeral abduction in children with suspected EDS, however, this study will analyze range of motion during functional tasks in both the glenohumeral and wrist. Normative pediatric data from existing research will be used to

compare the hEDS participants to a normal group. The results of this hypothesis can assist in understanding the phenotypic characteristics in children with hEDS.

*Aim 2*: To characterize pain location and severity in pediatric hEDS. This study will utilize the Numeric Rating Scale-11 to identify specific pain levels at multiple bodily landmarks (Miró, Castarlenas, & Huguet, 2009). It is hypothesized that children with hEDS will identify multiple locations of pain. It is also hypothesized that their highest level of self-reported pain will be greater than 4/10 on the Numeric Rating Scale-11 (NRS). A score above 4/10 on the NRS considered a moderate-to-severe pain level, identifying a level of pain in which a treatment intervention is often necessary (Miró et al., 2009). The findings of this hypothesis may identify common pain locations and severity in pediatric hEDS.

*Aim 3:* To understand common medical, clinical, or pharmaceutical treatments for pain management in pediatric hEDS and to identify the level of parent satisfaction for each treatment. It is hypothesized that each parent will report trialing multiple treatments to manage their child's hEDS pain and it is also hypothesized that the average level of satisfaction will be "Dissatisfied" on a Likert satisfaction scale (Albaum, 1997). Current research reports frequent dissatisfaction in treatment of hEDS in adults (M. C. Scheper et al., 2013). The information obtained from this aim will provide insight into the current treatment regimen for children with hEDS and whether the treatments utilized have benefits in relieving pain for the children. A non-standardized list of treatment methods developed by Dr. Joyce Engel and colleagues will be used with the parent of the participant to identify treatments that have been trialed to manage hEDS pain, the length of time the treatment has been utilized, and the parent's perception of satisfaction in using the treatment.

*Aim 4:* To understand how pain severity can impact the child's self-perception of disability due to pain. Pain severity will be measured through NRS while functional disability will be measured through the Functional Disability Inventory (FDI) (Walker & Green, 1991). It is hypothesized that children with higher ratings of pain will have higher scores of perceived disability in the Functional Disability Inventory. The information gathered from this hypothesis will assist in establishing the impact of hEDS on participation in ADL that are predominantly difficult for this population. Further, the information collected can provide specific information on the lived experience of children with hEDS and the aspects of occupational participation that are most difficult.

**Significance to Occupational Therapy**

The results of this study may offer clinical implications for occupational therapy practice. Children and adults with hEDS experience musculoskeletal problems that may identify a need for occupational therapy intervention. The data collected within this study regarding the glenohumeral kinematics during ADL in children with hEDs can inform an occupational therapist on common treatment interventions or symptoms in this population. Additionally, the pain assessment data obtained in this study may influence the intervention process for chronic pain management to emphasize teaching patients long-term coping mechanisms and ways to adapt engagement in occupations. Occupational therapy treatment would benefit the patient through pain management, treatment of musculoskeletal dysfunction, reduction of disability, and in educating patients on safe body mechanics.

## Literature Review

### *Anatomy and Kinematics of the Glenohumeral Joint*

The shoulder is a complex and very important joint within the body. Compiled of intricately placed ligaments, bones, and muscles, it allows for a wide range of motion for completing upper extremity activities of daily living. The shoulder provides three degrees of freedom consisting of flexion and extension, internal and external rotation, and adduction and abduction (Table 1). The shoulder provides the strength and positioning to support the arm, hand, and wrist positions for object manipulation and reaching (Neuman, 2017a).

The bones within the shoulder girdle create the four joints that comprise the entire shoulder. The glenohumeral joint is comprised of the humerus articulating with the glenoid fossa. The sternoclavicular joint is created by the proximal clavicle articulating with the sternum. At the distal clavicle, there is and connection with the acromion of the scapula that creates the acromioclavicular joint. The scapulothoracic joint is not considered a true joint as it is an interface between the thorax and the scapula (Neuman, 2017a).

The primary range of motion is found within the glenohumeral joint. In this joint, the glenoid fossa provides a shallow basin in which the large head of the humerus moves within. While this wide range of motion is beneficial for functional reaching tasks, it leaves the joint itself very unstable. To provide stability to the joint, the ligaments and muscles are position strategically to increase the support (Figure 1). Within the glenohumeral joints, there are three strong capsular ligaments. The superior glenohumeral ligament is responsible for resisting excessive external rotation and inferior or anterior movement of the humeral head. This ligament is taut when in anatomical position, however, it slackens during abduction above 40 degrees. The middle glenohumeral ligament primarily functions to restrict anterior movement of the humeral

head and excessive external rotation. It is involved in stabilizing the joint during most shoulder motions, though it is slack at the internal rotation position. Three primary components comprise the inferior glenohumeral ligament: the anterior band, the posterior band, and the axillary pouch. The anterior band is the strongest and thickest of the three and collaborates with the posterior band to restrict external and internal rotation. The axillary pouch holds the suspended humeral head in a cradle, adding more stability, while also resisting inferior and anterior humeral head movements. Outside of the capsular ligaments, lays the coracohumeral ligament. This ligament works to limit inferior translation and external rotation of the humeral head (Neuman, 2017a).

Because of the unstable structure of the shoulder complex, it has the ability to move in all three planes of motion while offering three degrees of freedom: abduction and adduction, flexion and extension, and internal and external rotation. Each movement at the glenohumeral joint will involve all four joints that comprise the shoulder. Abduction and adduction involve the humerus moving within the frontal place through the anterior-posterior axis of rotation. Typically, the human body allows for 120 degrees of abduction. It should be noted that the motion of abduction requires the upwards rotation of the scapula. If the upward rotators of the scapula are compromised, full abduction is not possible. Flexion and extension at the glenohumeral joint occur within the sagittal plane around the medial-lateral axis of rotation. Pure flexion has been found to be 0-120 degrees. However, flexion can occur up to 180 degrees with the involvement of the scapulothoracic joint. Shoulder extension can actively allow for 65 degrees of motion, though passively one may be able to reach around 80 degrees. Internal and external rotation can occur in an adducted or abducted position. In both of these positions, internal rotation yields the same range of motion value of 75-80 degrees. However, external rotation yields different values between two positions. When in adduction, the external rotation range can result in 60-79

degrees of motion. Conversely, when in abduction, external rotation can yield a full 90 degrees of motion (Neuman, 2017a).

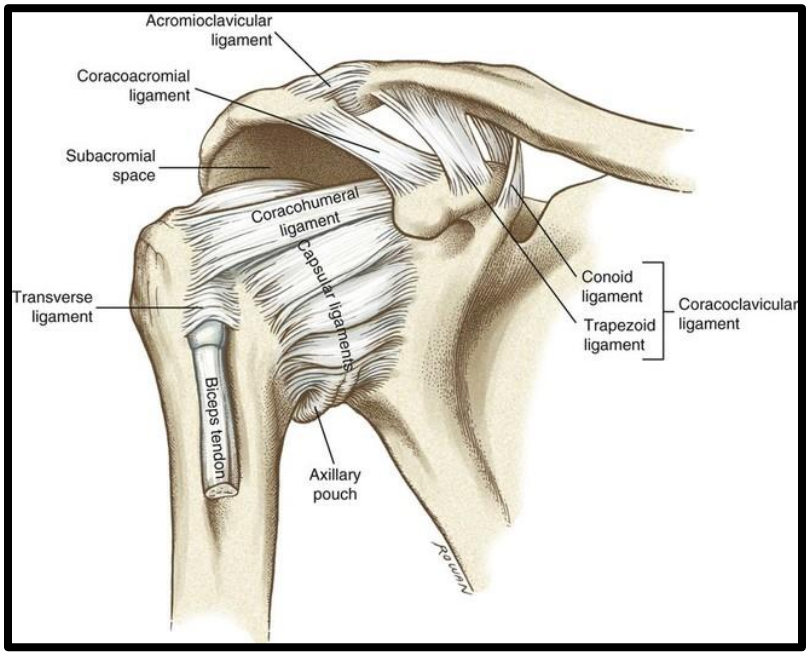



Figure 1: Glenohumeral Joint with Primary Ligaments

Table 1: Shoulder Range of Motion and Activities of Daily Living (ADL) (Neuman, 2017a)

| Shoulder Motion | Range of Motion (degrees) | Corresponding I/ADL |
|---|---|---|
| Flexion | 0-180 | Picking up an item from overhead, reaching up to wash or comb hair, donning a t-shirt |
| Extension | 0-65(80) | Reaching into a back pocket, performing toilet hygiene, |
| Abduction | 0-170 | Reaching out to the side for an item, combing hair, carrying items, donning clothes |
| Internal rotation | 0-75 | Reaching across midline, buckling seatbelt, brushing teeth, driving |
| External Rotation | 0-70 | Reaching into back pocket, combing hair, waving, driving |

*Anatomy and Kinematics of the Wrist Joint*

Though small, the wrist is an especially complex joint within the upper extremity. The wrist serves multiple purposes; however, its function is heavily involved in that of the hand and forearm during ADL. Without an efficiently and effectively functioning wrist, the hand and forearm would be of limited use and participating in self-cares would be especially difficult for any individual. The wrist offered two and arguably three degrees of freedom. In the sagittal plane, flexion and extension are the primary movements with flexion yielding 0-70 degrees and extension 0-60 degrees. In the frontal plane, ulnar deviation provides 0-35 degrees and radial deviation allows for 0-15 degrees of motion. Wrist circumduction is the full circular motion of the wrist, combining all four movements, however, this is not yet considered a true degree of freedom (Neuman, 2017b)

The wrist contains eight uniquely shaped carpal bones that are compacted into a small space to provide the wide variety of movements. The eight carpal bones contain the proximal row consisting of the scaphoid, lunate, triquetrum, and pisiform bones. Distally, the four bones are the trapezium, trapezoid, capitate, and the hamate. The proximal row of the carpal bones has been found to be more flexible allowing more freedom for movement, while the distal row is significantly more rigid in which the four distal bones will move as a singular unit. The carpal bones are connected to each other through intrinsic ligaments that are classified as short, intermediate, and long ligaments. The short ligaments are found at the distal row of the carpals both on the palmar and dorsal surfaces. There are many intermediate ligaments within the wrist. However, arguably the most important is the scapholunate ligament. This ligament stabilizes the lunate onto the scaphoid to allows cohesive movement between the two bones. Lastly, long ligaments consist of the palmar and dorsal intercarpal ligaments. The palmar intercarpal ligament

is more rigid and attaches at the palmar surface of the capitate. The dorsal intercarpal ligament provided transverse stability by connecting the trapezium, scaphoid, and triquetrum (Neuman, 2017b)

The proximal row of carpal bones connects to the forearm primarily through the articulation with the radius called the radiocarpal joint. The concavity of the radius in combination with the triangular fibrocartilage create the perfect fit into the convex carpal bones. Connecting the radius and ulna to the carpal bones is performed by the extrinsic ligaments of the wrist. The primary extrinsic ligaments are the dorsal radiocarpal ligament, the palmar radiocarpal ligaments and the triangular fibrocartilage complex (TFCC). Attaching at the dorsal distal radius, the dorsal radiocarpal ligament assists in guiding the wrist through its natural movement patterns. This ligament is especially important as it houses many mechanoreceptors to assist in wrist proprioception. The palmar radiocarpal ligaments are a group of ligaments that provide transverse support in connecting the radius to the palmar surfaces of the carpal bones. Lastly, the TFCC is found in the ulnocarpal space in which it provides a strong connection between the ulna and the radius, while also providing the necessary space for the radius to move freely during pronation and supination (Neuman, 2017b)

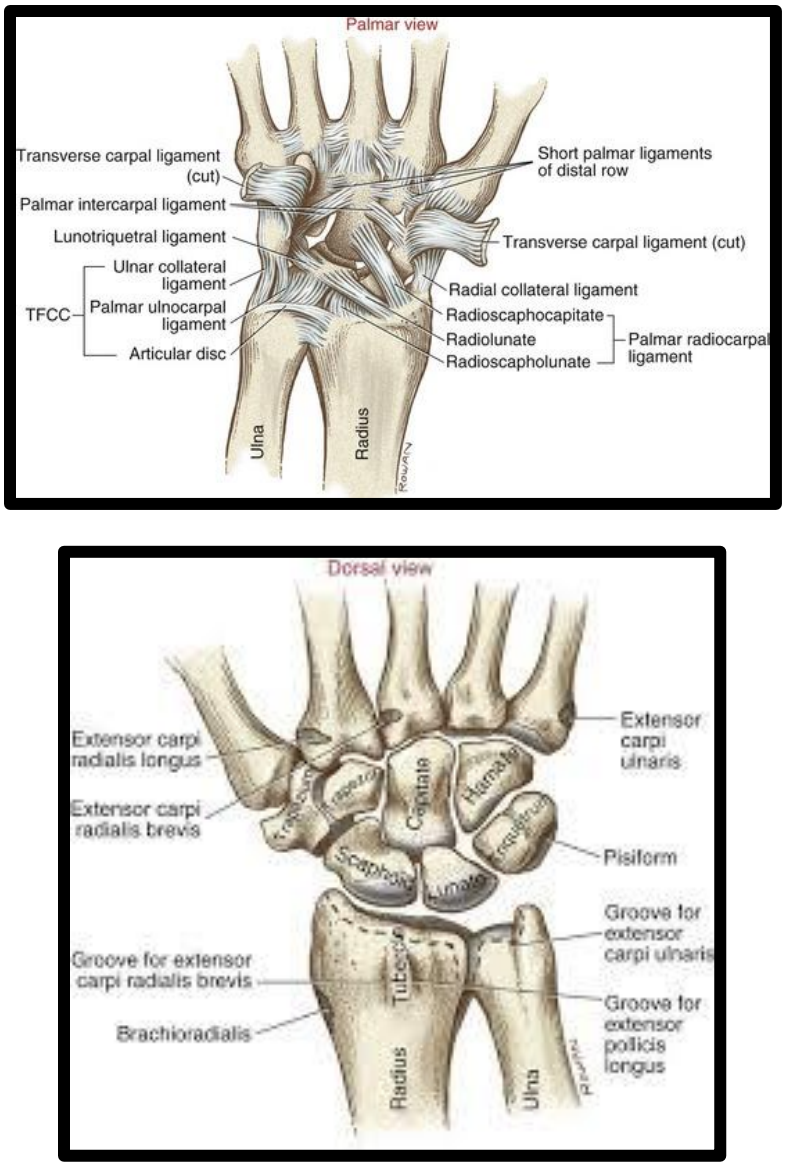



Figure 2: Palmar and Dorsal Ligaments of the Wrist (Neuman, 2017b)

Table 2: Wrist Range of Motion and Corresponding I/ADL (Neuman, 2017b)

| Wrist Motion | Range of Motion (degrees) | Corresponding I/ADL |
|---|---|---|
| Flexion | 0-70 | Functional grasp patterns, eating, dressing tasks |
| Extension | 0-60 | Functional grasp patterns, combing hair, brushing teeth, |
| Ulnar Deviation | 0-35 | Functional grasp patterns, typing, writing driving |
| Radial Deviation | 0-15 | Functional grasp patterns, typing, writing, eating, grooming tasks |

*Hypermobile Ehlers-Danlos Syndrome*

The Ehlers-Danlos Syndromes (EDS) consists of thirteen different subtypes of connective tissue disorders (Tinkle et al., 2017). Within the subtyped, hEDS is the most common, making up around 80% of the EDS population (Tinkle et al., 2017). Individuals with hEDS frequently present with joint hypermobility and slightly hyper-extensible skin (Levy, 2004). Joint hypermobility is classified as an individual's ability to actively or passively more their joints beyond a normal maximum range of motion (Colombi, Dordoni, Chiarelli, & Ritelli, 2015). Individuals with hEDS frequently find this condition difficult to manage, as chronic pain and fatigue are frequent symptoms in combination with hypermobile joints (Gurley-Green, 2001). The population of hEDS represents a high portion of patients seeking medical treatment for musculoskeletal problems and pain management (Johannessen, Reiten, Løvaas, Maeland, & Juul-Kristensen, 2016). However, without a genetic identifier, clinicians are unable to appropriately and accurately diagnose individuals with hEDS; leaving many patients struggling to find an appropriate diagnosis and treatment of their pain (Gurley-Green, 2001).

Symptoms of hEDS can change throughout the lifespan, making it especially difficult to diagnose at the pediatric level. As a result, children must withstand the brunt of this condition as the pain and fatigue interrupts their daily life. Participating in school and physical activities are often the greatest hurdles for children with hEDS. In a traditional school setting, many children are absent for a large portion of the school year due to the immense impact of the hEDS symptoms. However, other children  are forced to become home schooled to accommodate their need for more breaks and to work around their symptoms (Murray, Yashar, Uhlmann, Clauw, & Petty, 2013).

*Classification and Diagnostic Criteria*

The classification and diagnostic criteria of generalized joint hypermobility and EDS has evolved over time as the research surrounding these conditions have too evolved. Early studies of joint laxity were classified with five criteria that included passive hyperextension of the fingers, elbows, knees, dorsiflexion, and eversion of the foot. This criteria yielded a 7% prevalence of joint hypermobility in a sample of 285 children (Carter & Wilkinson, 1964). Nearly twenty years later, an international nosology was created for heritable connective tissue disorders (HCTDs). this nosology included only nine subtypes within the Ehlers Danlos Syndromes. The hypermobility type of EDS was included within this nosology which reported the primary manifestations as articular hypermobility, dermal hyper-extensibility, and minimal scarring. However, this nosology was greatly critiqued as specific locations and degrees of hypermobility were not specified, resulting in an ambiguous diagnostic criteria (Beighton et al., 1997). Ten years after the HCTD nosology, another updated version was published. This version utilized the Beighton Scale, a measure of hypermobility, and all established genetic discoveries to advance the classification process of EDS (Beighton et al., 1997). The nosology classified six major subtypes of EDS, with all subtypes consisting of specific major and minor criteria, increasing the specificity of the EDS diagnoses. Though, it was not for another twenty years until Malfait et al. (2017) established the current classification of EDS, making the diagnostic criteria for hEDS even more reliable and detailed. The updated criteria include all thirteen different subtypes of EDS. The primary requirement of hEDS in these criteria is the presence of generalized joint hypermobility. These criteria require joint hypermobility to be measured using the Beighton scale, which requires patients to score greater than or equal to 5/9 to be considered hypermobile. This scale measures hypermobility within the thumb, wrist, knees, and elbows.

(Malfait et al., 2017; Smits-Engelsman, Klerks, & Kirby, 2011). Additional criteria involves the possible presence of velvety skin, chronic widespread pain, recurrent joint dislocations, and the exclusion of all heritable connective tissue disorders (Levy, 2004).

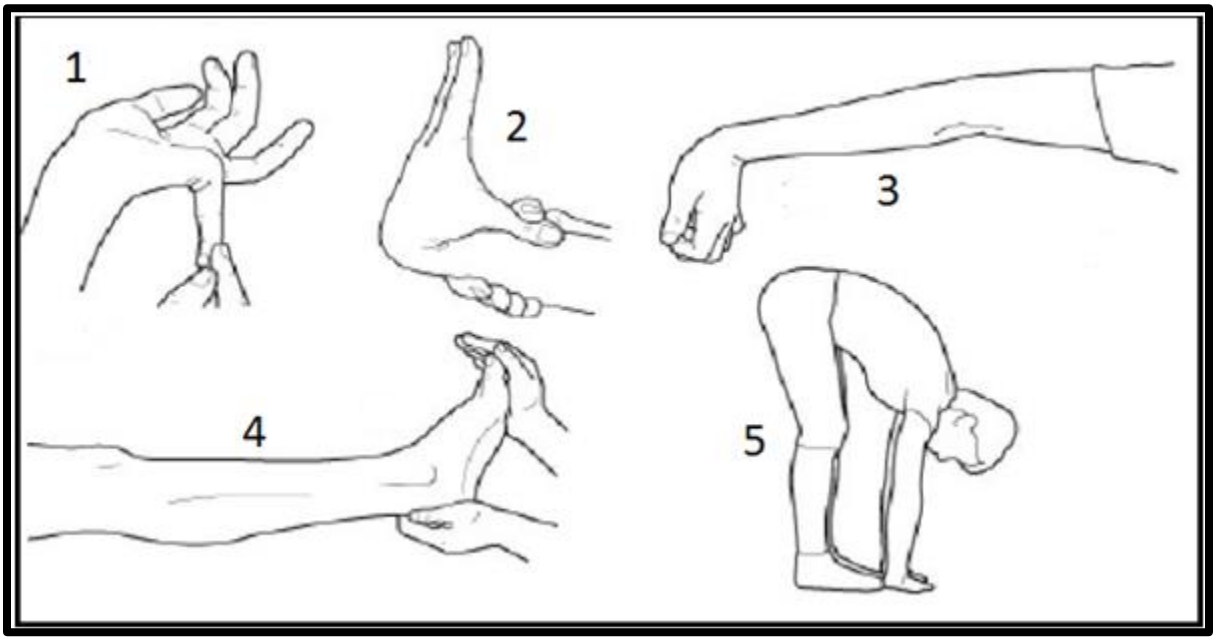



Figure 3: Beighton Criteria (Beighton score, Physiopedia)

*Musculoskeletal Manifestations in Pediatric Hypermobile Ehlers-Danlos Syndrome*

Within the pediatric population of hEDS, the primary musculoskeletal manifestation consists of ligamentous laxity and hypermobile joints. The ligamentous laxity can be so extreme that frequent joint subluxations and dislocations occur spontaneously, even when repositioning while sleeping (Gazit, Jacob, & Grahame, 2016). In pediatric hEDS, joint dislocations have bene reported to be the most common at the shoulder, ankle, temporomandibular joint, and knees (Castori, 2016; De Coster, Martens, & De Paepe, 2005; Johannessen et al., 2016). As a result of these frequent joint dislocations, children with hEDS, often experience shoulder discomfort or experience pain as if they have twisted an ankle or had their knees give out, and

temporomandibular syndrome (Hagberg, Berglund, Korpe, & Anderson-Norinder, 2004; Levy, 2004). While the pain of the joint instability can be primary concern at the time of dislocation, the long-term effect can pose more significant medical problems. Early onset of osteoarthritis has shown to be a common diagnosis in pediatric hEDS (Levy, 2004). Further, the frequent joint trauma will result in ongoing tendinitis and bursitis (Lies Rombaut et al., 2011). The ongoing experience of these joint traumas and pain can ignite Kinesiophobia, a fear of movement, at a young age in children with hEDS, leaving them fearful to participate in any exercise of physical activity (Kazkaz & Grahame, 2018; M. Scheper, de Vries, Verbunt, & Engelbert, 2015).

Hypermobility within the shoulder and wrist can be common within pediatric hEDS. Frequent dislocations at the shoulder can cause nerve impingement resulting in acute and chronic pain throughout the extremity (Johannessen et al., 2016). Children with hEDS are especially impacted by pain at the wrist, as writing and typing can induce pain, impacting their school participation (Chopra et al., 2017). The shoulder and wrist are frequently if not always utilized in the participation of ADL. While in a healthy population this would be appropriate, children with hEDS may be constantly experiencing microtraumas at both joints every day as they participate in their self-cares. While a sudden impact may not be noticeable at the time of these microtraumas, over time the child may be progressively damaging their joints. Further, the child may learn compensatory movement patterns to avoid pain and to limit their active range of motion at the joints to reduce the chance for joint trauma (Syx, De Wandele, Rombaut, & Malfait, 2017). While this may seem as a solution to protect the joints, these compensatory movements are equally detrimental, as muscle groups in the upper extremity can become weak, imbalanced, and may result in further nerve impingement due to the improper positioning of the joints (Camerota, Celletti, Castori, Grammatico, & Padua, 2011).

*Pain and Psychological Manifestations in Pediatric Hypermobile Ehlers-Danlos Syndrome*

Within pediatric hEDS, pain and psychological manifestations can have the most pernicious impact (Hagberg et al., 2004; L. Rombaut et al., 2014), the implications of hEDS can induce secondary diagnoses of Major Depressive Disorder (MDD) and Generalized Anxiety Disorder (GAD) in combination with chronic pain and fatigue (Castori, 2016; Hagberg et al., 2004; L. Rombaut et al., 2014). Nearly 75% of the pediatric population of hEDS reports chronic pain symptoms by the age of 15 (Gazit et al., 2016).

The impact of chronic pain is vast as the children with hEDS have reported moderate to severe musculoskeletal and neuropathic pain experiences every day, with some reporting that their pain is constant (Voermans, Knoop, Bleijenberg, & van Engelen, 2011). Musculoskeletal pain most frequently occurs at the joints, causing reports of stiff and aching joints (L. Rombaut et al., 2014). Neuropathic pain can result in an experience of shooting or burning pain in combination with numbness in some areas (Camerota et al., 2011). Those with early onset osteoarthritis report a more severe form of aching joints and sharp pain; however, this type of pain is known to be exacerbated by activity (Levy, 2004). Each child with hEDS may experience their pain differently, however, the debilitating pain often starts early in adolescent years, and can even worsen throughout the lifespan (Sacheti et al., 1997).

Myalgic Encephalomyelitis or Chronic Fatigue Syndrome (ME/CFS) is the experience of immense fatigue that does not improve with rest. For children to obtain a diagnosis of ME/CFS, the fatigue is consistent for more than six months ((CDC), 2020). Because hEDS is difficult to diagnose, ME/CFS is often the first diagnosis an individual will receive (Hakim, De Wandele, O'Callaghan, Pocinki, & Rowe, 2017). Children with ME/CFS are greatly inhibited in their ability to participate in a typical school setting, ADL, and leisure or play activities. Reports on

lived experiences of hEDS identify the detriment that chronic fatigue can bring. The fatigue can be so overwhelming, individuals with ME/CFS will spend their day sedentary, unable to leave their bed, and are then subjected to sleepless nights (CCHMC, 2014; Hakim et al., 2017).

Generalized Anxiety Disorder (GAD) and Major Depressive Disorder are frequently reported in both the adult and pediatric populations of hEDS. These conditions often develop within adolescence and continue to impact the individual into adulthood. A singular cause of both GAD and MDD is unknown. However, the impact of experiencing fear or nervousness of not knowing when their pain with stop, lack of social participation due to pain, and Kinesiophobia after suffering joint trauma can be major factors in provoking these secondary conditions. Further, many individuals with hEDS are not properly diagnosed until adulthood. This can cause immense stress in adolescence as the child is left battling a hidden condition, one in which their own family may not fully understand or believe in (Gazit et al., 2016).

*Treatment of Pain and Musculoskeletal Dysfunction Pediatric Hypermobile Ehlers-Danlos Syndrome*

The lack of accurate and appropriate diagnostic criteria for patients with hEDS has then negatively impacted the efficacy in treating patients with this condition. Frustrations regarding treatment for pain and musculoskeletal dysfunction is often cited in research. Children and adolescence with hEDS may not obtain an appropriate diagnosis of hEDS until well into their adulthood, leaving then with years of unmanaged and unruly symptoms. During the time without a diagnosis, the child may be enduring chronic pain and ME/CFS while developing psychological conditions as a result (CCHMC, 2014). Further, parents of these children are left without answers, requiring them to trial their own remedies to alleviate the symptoms. Once a

diagnosis is formally provided, treatment for this condition are often reports insufficient and may leave an individual unsatisfied (Lies Rombaut et al., 2011).

While there is not a singular treatment protocol for individuals with hEDS, research supports the importance of providing multidisciplinary care to address all aspects of the symptoms of hEDS (Gazit et al., 2016; M. C. Scheper et al., 2013). The multidisciplinary care of hEDS often includes rheumatologists, physical and occupational therapists, and psychologists. The primary goals of treatment for hEDS should involve preventing physical deterioration, pain management, and optimizing functional abilities and participation (Hakim et al., 2017). Physical therapists can provide education and rehabilitation focused on joint protection strategies and injury prevention. While occupational therapists can provide treatment for pain management, improved proprioception and can fabricate splints for handwriting (Johannessen et al., 2016; Keer & Butler, 2010; Levy, 2004). Outside of physical rehabilitation the rheumatologist and psychologist can address the systemic and psychological symptoms of hEDS with treatment and pharmaceutical intervention when appropriate. Successful multidisciplinary treatment involves overall patience, strong communication with the patient and their family, and sensitivity to the patient's symptoms (CCHMC, 2014). Above all, it is important that providers understand the importance for hEDS patients to feel a sense of control over their condition (Gurley-Green, 2001).

*Impact of Hypermobile Ehlers-Danlos Syndrome on Occupational Performance and Quality of Life in Pediatric Hypermobile Ehlers-Danlos Syndrome*

The lack of diagnostic efficacy in combination with both physical and psychological manifestations contribute to decreased quality of life and occupational engagement in both adults and children with hEDS (CCHMC, 2014; De Wandele et al., 2013). The years without a

diagnosis are spent visiting a series of doctors and specialists, requiring an excessive amount of both time and money before help can be provided (Castori, 2016). Further, at times, hEDS can be an "invisible" condition, in which the symptoms are not overt to others. This aspect can cause emotional divide between individuals with hEDS and their loves ones, as family and peers may not fully believe the severity of the individual's symptoms (Gurley-Green, 2001).

The manifestations of hEDS can have major negative impacts on the patient's occupational performance. Occupational performance consists of the individual's participation in their activities of daily life and instrumental activities of daily life (AOTA, 2020). The impact of the chronic pain, anxiety, depression, and musculoskeletal dysfunction all contribute to the lack of occupational engagement of the individual with hEDS (M. Scheper et al., 2015). For individuals who are severely affected with this condition, every day is a struggle to participate in their own lives. Simple every day activities can pose to be too much to bear. It is often reported that patients with hEDS find it difficult to remain employed and children are forced to miss days of school as the pain and fatigue requires the individual to rest more frequently. When joint pain is experienced within the hands and fingers, some children are unable to even write; as the movements of writing greatly exacerbates the pain. Family and friend relationships are also negatively impacted as the individual with hEDS may not be able to participate in family activities or events (Gurley-Green, 2001)

*Numerical Rating Scale*

The eleven-point Numerical Rating Scale (NRS) is frequently used both within clinical and research settings for measuring pain intensity (Miró et al., 2009). The NRS provides a quick way to get a measure for the patient's subjective experience of his or her pain intensity (Breivik et al., 2008). When measuring an individual's level of pain, understanding the pain intensity is

one of the most crucial components and clinically relevant dimension of pain (Hjermstad et al., 2011). This pain scale asks the patient to grade his or her pain from 0 to 10. On this scale, a choice of zero would indicate no pain at all, while an answer of ten would indicate the worst pain imaginable (Bailey, Daoust, Doyon-Trottier, Dauphin-Pierre, & Gravel, 2010). Score interpretation of the NRS provides three categories of pain levels (Breivik et al., 2008). Scoring zero indicates the absence of any pain. Individuals scoring between one and three will indicate mild pain, while those who score between four and six experience moderate pain. Those who designate seven to ten for their pain are experiencing severe pain (Breivik et al., 2008). The questions regarding pain are frequently asked in the context of right now, within the past twenty-four hours, and within the past week (Breivik et al., 2008). The NRS is easy to understand, translate, and is often the preferred method of quantifying pain intensity within patients (Hjermstad et al., 2011). While this tool is commonly used and researched in the adult population, it has been found to be of appropriate use within pediatrics as well (Bailey et al., 2010; Hjermstad et al., 2011; Miró et al., 2009). Research has been conducted regarding the validity of the NRS within the pediatric population and has shown that the NRS is appropriate to use with children and adolescents. Studies have shown strong validity within the NRS as well as strong construct, content, and discriminant validity. Test-retest of the NRS was not well-established, however, this may be due to the rapid changing of pain and the individual's ability to easily remember (Bailey et al., 2010; Hjermstad et al., 2011; Miró et al., 2009).

*Functional Disability Inventory*

The Functional Disability Inventory (FDI) is a widely used measure that yields a quantitative score of impairment within children and adolescents experiencing chronic pain. This measure has been used with children experiencing fibromyalgia, abdominal pain, and

musculoskeletal pain symptoms (Kashikar-Zuck et al., 2011). This tool is a primary measure utilized in clinical trials for pediatric chronic pain and has been recommended by the PedIMMPACT for assessing physical functioning in clinical trials. The FDI can be utilized as a self-assessment, with language that adheres to the reading level of children at the age of 8 or can be administered interview style. The FDI requires roughly ten minutes to administer, though this varies depending on the reading skills of the child if being administered as a self-assessment, and only five minutes to score. No special training or certification is required to administer this tool. This tool assessed the level of difficulty the individual experiences while completing a variety of daily tasks within their home and at school. The items within the FDI were established by reviewing items from adult impairment measures and adapting them to adhere to the pediatric population. Pilot testing was initially conducted to ensure that the items suited the lives and experiences of children, and changes were made accordingly to adapt to the children. The items within the FDI include doing chores, being at school all day, and walking upstairs. The child or adolescent then rates each activity utilizing the five-point Likert scale with a score of zero indicating no trouble in completing the activity and a score of four indicating an impossible task for them to complete. The total score of the finished FDI will indicate the level of perceived disability due to pain of the individual. The higher the total score the greater indication of pain related disability (Flowers & Kashikar-Zuck, 2011). A score of less than 12 will identify no or minimal disability, a score of 13-29 identifies moderate disability, and a score of greater than or equal to 30 identifies severe disability due to pain. Moderate disability has been found in most patients with chronic disability, while the severe disability group frequently experiences a higher rate of pain and depressive symptoms. Very few individuals will display scores of zero or 60, with no current floor or ceiling affect yet identified. Typically, children with chronic pain will

fall within the moderate to severe categories of disability due to pain. Healthy children with no reported chronic pain will fall below or minimal disability with scores typically ranging from 3-8 (Flowers & Kashikar-Zuck, 2011; Kashikar-Zuck et al., 2011).

The FDI is an efficient and user-friendly tool to use for measuring and tracking the disability due to chronic pain in children and adolescents. Multiple studies have been conducted to test the validity and reliability of this measure. Kashikar-Zuck (2011) identifies the FDI with high internal consistency, moderate to high test-retest reliability, moderate cross-informant (parent-child) reliability, and good predictive validity. Flowers et al., identified the FDI to have internal consistency, stability, content validity, criterion validity, ability to detect change, and strong psychometric properties.

*Vicon Motion Capture System*

Kinematics is the way in which we are able to describe body motions without including information involving forces or torque generated by the muscles of the body. This provides information into the planes of motion, ranges of motion, and axis of rotation, that a joint or a body segment is moving during a task. The human body moves within three planes of motion: sagittal, frontal, transverse. When moving within these planes, each joint or body segment is also rotating around an axis of rotation. Measuring joint kinematics with 3-dimensional motion capture software provides information regarding the entire joint motion within all planes and around all axis of rotation (Neuman, 2017a). To obtain 3-dimensional kinematic data using motion capture, reflective markers are first placed on the participant on specific bodily landmarks. These markers are recognized by the motion capture cameras, and their position and movements are measured and recorded. This study utilizes a Vicon Motion Capture System (Oxford Metrics) that includes 15 motion cameras that surround the participant when completing

a task. The reflective markers on the body need to be seen by two cameras in order to recognize the reflective marker position on the body. Data is then collected at a frequency of 120 frames per second (Hz), which provides detailed information on the movements of the body and allows us to recognize if any reflective markers are missing within a frame. Once the data is collected, the reflective markers are then labeled with the designated landmarks. This allows us to then compare the location of one landmark to another providing us with information regarding the specified joint and how it moves while completing a task.

# II. MANUSCRIPT

## Introduction

Ehlers-Danlos syndrome (EDS) is a group of heritable connective tissue disorders, consisting of thirteen different subtypes. Among the thirteen, Hypermobile Ehlers Danlos Syndrome (hEDS) is the most common. Individuals with this condition present with frequent joint instability that results in ongoing subluxations and dislocations. Secondary diagnoses of this condition include chronic pain, Myalgic Encephalomyelitis/Chronic Fatigue Syndrome (ME/CFS), Major Depressive Disorder (MDD), and Generalized Anxiety Disorder (GAD). The diagnostic process of hEDS is convoluted by the lack of a genetic identifier (Malfait et al., 2017). Individuals with hEDS are often not diagnosed until later in adulthood, leaving their adolescent life full of confusion and difficulty in managing the symptoms (Engelbert et al., 2017). As a result, children with hEDS endure a childhood of difficulty in participating in school, activities, and engaging with peers.

The primary purpose of this study is to characterize pediatric hEDS to enhance the diagnostic process at the adolescent stage. This study analyzes pediatric upper extremity kinematics and pain outcomes to achieve insight into pediatric hEDS. The first aim of this study

is to characterize upper extremity 3D kinematics during components of activities of daily living (ADL). It is hypothesized that individuals with hEDS with have a greater range of motion during the tasks when compared to existing kinematic literature. The second aim focuses on characterizing pain locations and severity of children with hEDS. It is hypothesized that children with hEDS will identify multiple locations of pain and that their highest level of pain will be greater than 4/10 on the Numeric Rating Scale (NRS). The third aim of this study is to understand common medical, clinical, or pharmaceutical treatments for pain management in pediatric hEDS. It is hypothesized that each parent will report trialing multiple treatments to manage their child's pain and that their average level of satisfaction will be "Dissatisfied" on a Likert satisfaction scale. The fourth and final aim is to understand the impact of pain severity on the child's self-perception of disability due to pain. It is hypothesized that children with higher ratings of pain will have higher score on the Functional Disability Inventory.

Table 3: Aims and Hypotheses

| Aim | | Hypotheses |
|---|---|---|
| 1 | To characterize upper extremity 3D kinematics during activities of daily living in pediatric hEDS. | It is hypothesized that during the ADL, children with hEDS will display greater glenohumeral, wrist, and thoracohumeral range of motion when compared to existing kinematic literature. |
| 2 | To characterize pain location and severity in pediatric hEDS | A: It is hypothesized that children with hEDS will identify multiple locations of pain. |
| | | B: It is hypothesized that their highest level of self-reported pain will be greater than 4/10 on the Numeric Rating Scale-11 (NRS). |
| 3 | To understand common medical, clinical, or pharmaceutical treatments for pain management in pediatric hEDS and to identify the level of parent satisfaction for each treatment | A: It is hypothesized that each parent will report trialing multiple treatments to manage their child's hEDS pain |
| | | B: It is hypothesized that the average level of satisfaction will be "Dissatisfied" on a Likert satisfaction scale |
| 4 | To understand how pain severity can impact the child's self-perception of disability due to pain. | It is hypothesized that children with higher ratings of pain will have higher scores of perceived disability in the Functional disability inventory |

## Methods

This study was conducted at the University of Wisconsin-Milwaukee Mobility Lab in collaboration with Children's Hospital of Wisconsin Genetics Center. All study procedures were approved by the University of Wisconsin-Milwaukee Institutional Review Board. Prior to any data collection, participation and their parents provided informed consent to participate in this study. There were minimal risks associated with this study such as: skin irritation from the reflective marker set, or fatigue from repetitive movements during the 3D motion capture process. The risks were minimized by allowing each participant to take frequent breaks and providing proper training of the research team for all data collection components.

## Sample

This study consisted of eleven children with hEDS and their legal guardian. The participants were primarily female (6/11) with the mean age of the children being 13 (± 3 years) (Table 4). All children within this study have a diagnosis or suspected diagnosis for hEDS and were recruited through the Children's Hospital of Wisconsin Genetics Center. The inclusion and exclusion criteria for this study were based on the diagnostic criteria identified in the most recent international classification (Malfait et al., 2017).

Table 4: Participant Demographics

| Subject | Sex | Age | Beighton Score |
|---|---|---|---|
| 1 | Female | 15 | 6/9 |
| 2 | Male | 12 | 6/9 |
| 3 | Female | 14 | 5/9 |
| 4 | Male | 10 | 2/9 |
| 5 | Female | 15 | 6/9 |
| 6 | Female | 17 | 4/9 |
| 7 | Male | 17 | X |

| | | | |
|---|---|---|---|
| 8 | Female | 9 | 4/9 |
| 9 | Female | 8 | 5/9 |
| 10 | Male | 12 | 4/9 |
| 11 | Male | 14 | 2/9 |
| Average | | 13± 3.07 | |

Table 5: Inclusion and Exclusion Criteria for Recruitment

| **Inclusion Criteria** |
|---|
| - 8-18 years old, but not including 18 years of age<br>- Beighton score equal or greater than 5 out of 9<br>- No other diagnosis<br>- Symptoms of autonomic dysfunction, including: Postural orthostatic tachycardia syndrome (POTS), gastroparesis, and/or abnormal Quantitative Sudomotor Axon Reflex test (QSMART)<br>- Participation in the separate, current study at Children's Hospital of Wisconsin Genetic Center led by Dr. Basel |
| **Exclusion Criteria** |
| - Bone marrow transplant<br>- Inability or unwillingness on the individual (or parent/legal guardian) to provide clinical or family history<br>- Non-English speaking |

**Data Collection**

Data collection for this study was conducted at the University of Wisconsin-Milwaukee Mobility Lab. Participants were required to attend two separate days of data collection to decrease the risk of fatigue for the children with hEDS. The first day of data collection consisted of completing the Pain Location and Severity Questionnaire, Functional Disability Inventory (FDI) , and Treatment Questionnaire with both the child and their legal guardian. The second day of data collection consisted of collecting kinematic data through the use of Vicon Motion Capture System.

*Kinematic Data Collection*

All kinematic data were collected using the Vicon T-series motion capture system (Oxford Metrics). Each trial was collected at 120 Hz using a 15-camera 3D system. Retro reflective markers were placed along the upper and lower extremity bony prominences on each child. The children were then required to complete five trials of four upper extremity tasks. The movements in each task simulate components of ADL tasks. Starting position for each task consisted of the child standing with his or her hands relaxed at their side. All of the children in this study were encouraged to take rest breaks as needed throughout the data collection.

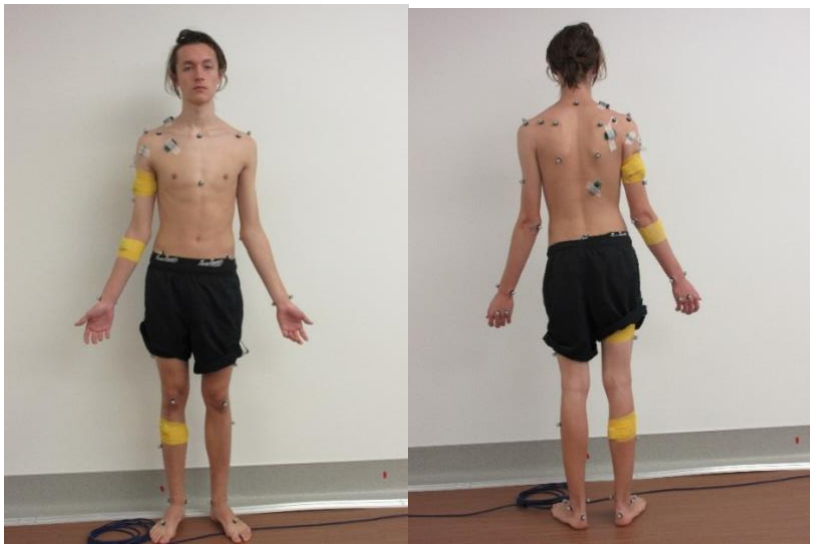


Figure 4: Participant with Retroreflective Marker Placement

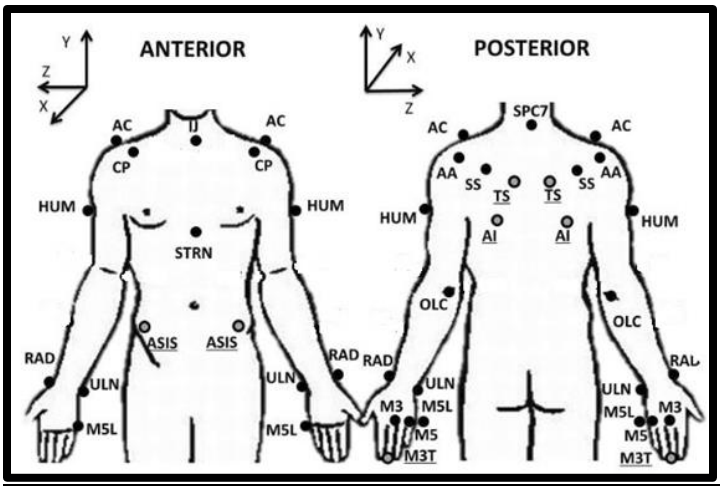



Figure 5: Custom 3D Bilateral UE Biomechanical Model (Schnorenberg et al., 2014)

Table 6: UE Marker List and Locations

| Segment | Marker | Location |
|---|---|---|
| Trunk | SPC7 | Spinous process, C7 |
| | STRN | Sternum, xiphoid process |
| | IJ | Incisura jugularis (suprasternal notch) |
| Clavicle | AC | Acromioclavicular joint |
| Scapula | AA | Acromial angle |
| | SS | Scapular spine, halfway between TS and AA |
| | TS | Trigonum Spine |
| | AI | Inferior Angle |
| | CP | Coracoid process |
| Humerus | HUM | Humerus |
| | OLC | Olecranon |
| Forearm | RAD | Radial Styloid |
| | ULN | Ulnar Styloid |
| Hand | M3 | Third Metacarpal |
| | M5 | Fifth Metacarpal |

Drinking Task

Prior to completing the trials, a table was placed in front of the child and was set to half the child's height. The child was then instructed to reach, using their dominant hand, for a water bottle that was placed on the table, simulate bringing the water bottle to his or her mouth to take

a drink, and then place the water bottle back onto the table. The task was completed when the child returned his or her hands to their side.

Figure 6: Participant Completing Drinking Task with Vicon Joint Segment Overlay

Combing Task

The combing task began with the child holding a comb in his or her dominant hand. The child was then instructed to simulate combing his or her hair once and then return their hands back to his or her side.

Reaching Across Midline Task

For the reaching across midline task, the child was instructed to use his or her dominant hand to reach across midline to his or her contralateral shoulder and then return his or her hands to the side.

Reaching Back to Ipsilateral Side Task

For the reaching back task, the child was instructed to use his or her dominant hand to reach back to their ipsilateral side and return his or her hands to the side.

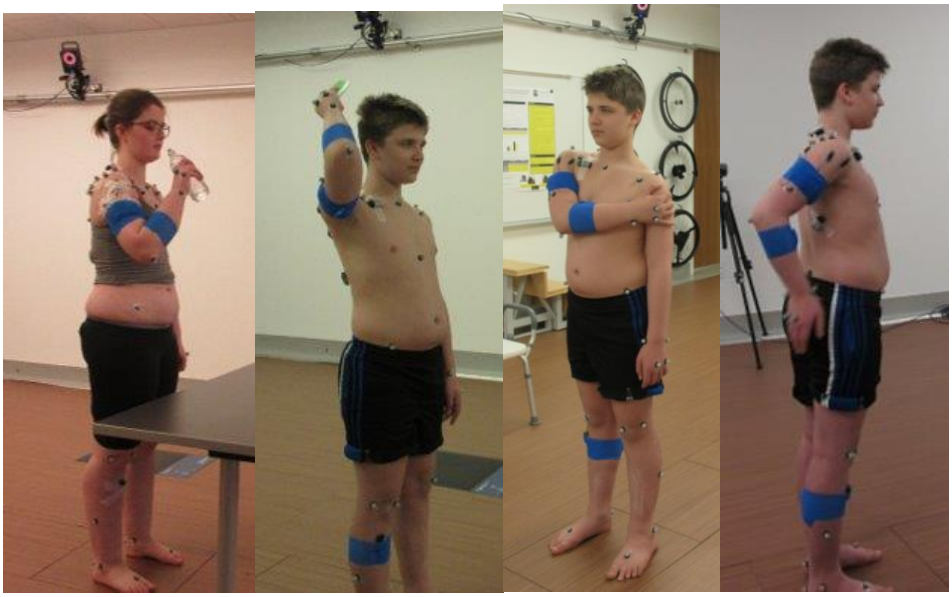
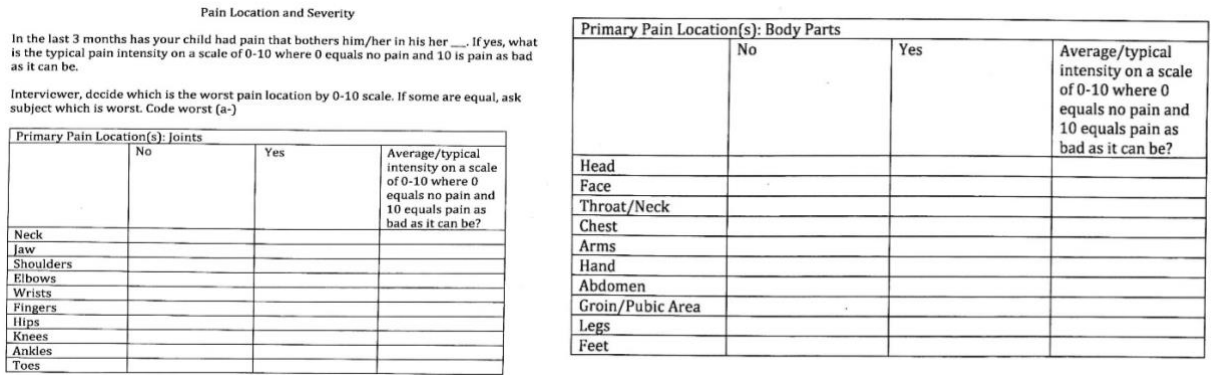


Figure 7: Participants Completing ADL Tasks

*Pain Location and Severity*

The Pain Location and Severity Questionnaire was developed by Dr. Joyce Engel to identify specific pain regions and severity in children with hEDS similar to previous studies (Sacheti et al., 1997) (Figure 7). The questionnaire assesses ten joint locations and ten bodily regions for pain presence and severity. The questionnaire was completed in a private quiet room with limited distractions. The interviewer verbalized the question listed at the top of the questionnaire and read aloud each joint and bodily location to the individual. The child/legal guardian would first answer 'yes' or 'no' to each joint or bodily location. If the child/legal guardian answered 'yes' then the interviewer would then ask them to rate their pain using the Numerical Rating Scale (NRS). A printed visualization was provided to each subject with the NRS scale to ensure accuracy in reporting. The interviewer would then scribe the ratings for each location. If the child/legal guardian wavered between two numbers, the interviewer would instruct the individual to pick the one number that best described the level of pain.



Pain Location and Severity

In the last 3 months has your child had pain that bothers him/her in his her ___. If yes, what is the typical pain intensity on a scale of 0-10 where 0 equals no pain and 10 is pain as bad as it can be.

Interviewer, decide which is the worst pain location by 0-10 scale. If some are equal, ask subject which is worst. Code worst (a-)

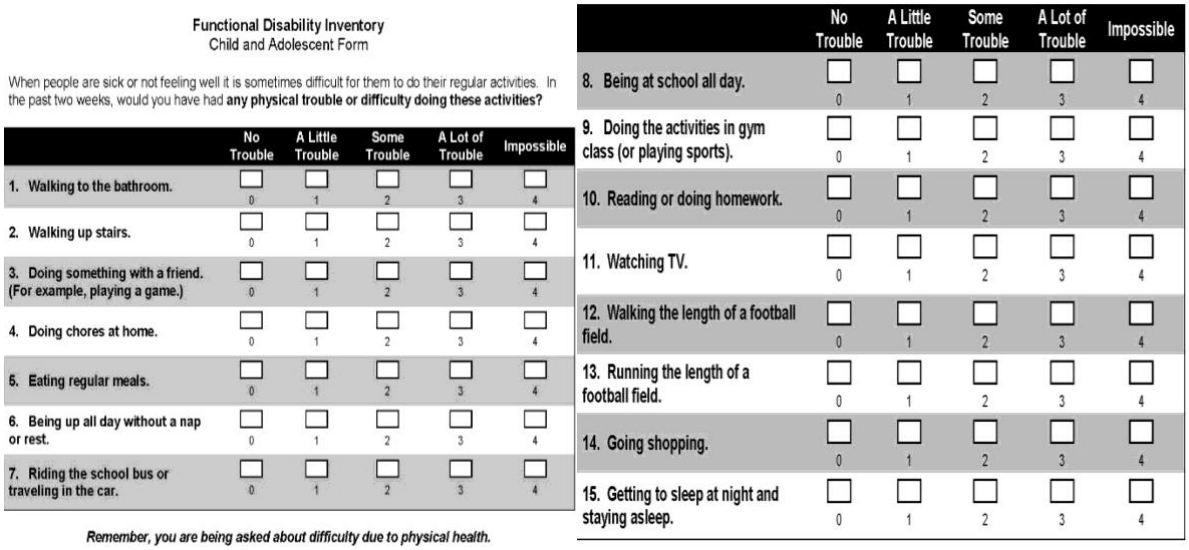
| Primary Pain Location(s): Joints | | | |
|---|---|---|---|
| | No | Yes | Average/typical intensity on a scale of 0-10 where 0 equals no pain and 10 equals pain as bad as it can be? |
| Neck | | | |
| Jaw | | | |
| Shoulders | | | |
| Elbows | | | |
| Wrists | | | |
| Fingers | | | |
| Hips | | | |
| Knees | | | |
| Ankles | | | |
| Toes | | | |

| Primary Pain Location(s): Body Parts | | | |
|---|---|---|---|
| | No | Yes | Average/typical intensity on a scale of 0-10 where 0 equals no pain and 10 equals pain as bad as it can be? |
| Head | | | |
| Face | | | |
| Throat/Neck | | | |
| Chest | | | |
| Arms | | | |
| Hand | | | |
| Abdomen | | | |
| Groin/Pubic Area | | | |
| Legs | | | |
| Feet | | | |

Figure 8: Pain Location and Severity Questionnaire

*Functional Disability Inventory*

The FDI is developed specifically for pediatric use. It quantifies the child's perceived level of disability due to pain and provides a parent proxy form for comparison (Kashikar-Zuck et al., 2011) (Figure 8). Similar to the Pain Location and Severity Questionnaire, the FDI was completed interview style in a quiet and private room. The child and their legal guardian were interviewed separately. Prior to completing the FDI, the interviewer read aloud the statement and question at the top of the page and provided the child/legal guardian with a visual representation of the Likert scale on the FDI. The interviewer verbalized each activity listed on the form and scribed the child/legal guardian's answer. If the child/legal guardian was unsure about their answer, the interviewer encouraged them to select the answer that best described their level of difficulty in completing the activity.



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?**

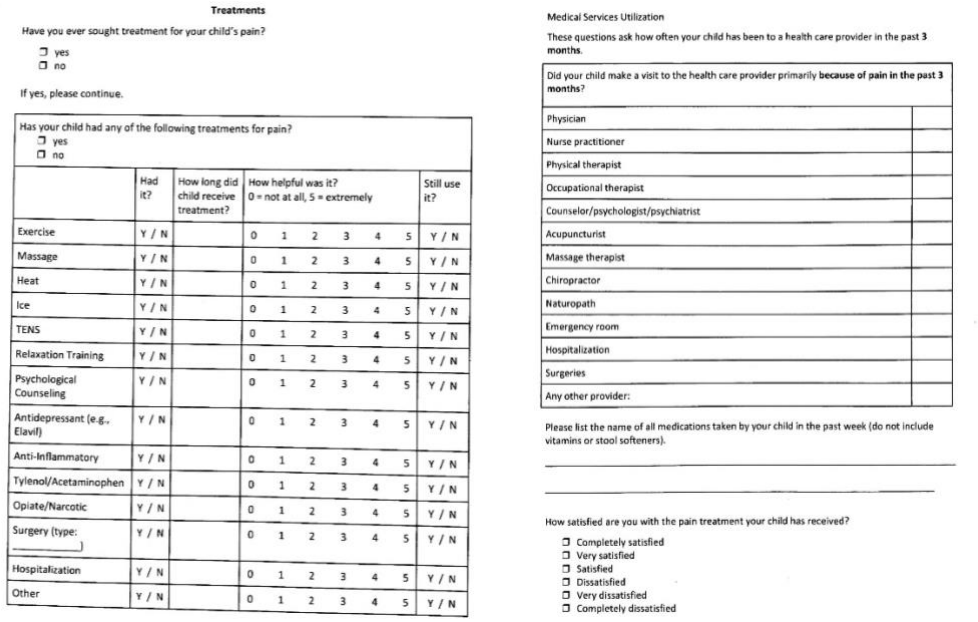
| | No Trouble | A Little Trouble | Some Trouble | A Lot of Trouble | Impossible |
|---|---|---|---|---|---|
| 1. Walking to the bathroom. | ☐ 0 | ☐ 1 | ☐ 2 | ☐ 3 | ☐ 4 |
| 2. Walking up stairs. | ☐ 0 | ☐ 1 | ☐ 2 | ☐ 3 | ☐ 4 |
| 3. Doing something with a friend. (For example, playing a game.) | ☐ 0 | ☐ 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 |
| 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 |

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

Figure 9: Functional Disability Inventory Child and Adolescent Form

*Treatment History Questionnaire*

The Treatment History Questionnaire was developed by Dr. Joyce Engel for this study to identify successful treatments within the pediatric hEDS population. This questionnaire was completed interview style with the legal guardian of the child with hEDS (Figure 9). The questionnaire collected a variety of information including specific treatments, frequency of treatment, medical services, and success level of each treatment. The interviewer aspect of the questionnaire aloud to the legal guardian. If the parent had questions at any time, the interviewer encouraged the legal guardian to provide the most accurate information related to his or her child's treatments.



**Treatments**

Have you ever sought treatment for your child's pain?

❒ yes
❒ no

If yes, please continue.

| Has your child had any of the following treatments for pain? ❒ yes ❒ no | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|
| | Had it? | How long did child receive treatment? | How helpful was it? 0 = not at all, 5 = extremely | | | | | | Still use it? |
| Exercise | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | Y / N |
| Massage | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | Y / N |
| Heat | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | Y / N |
| Ice | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | Y / N |
| TENS | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | Y / N |
| Relaxation Training | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | Y / N |
| Psychological Counseling | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | Y / N |
| Antidepressant (e.g., Elavil) | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | Y / N |
| Anti-Inflammatory | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | Y / N |
| Tylenol/Acetaminophen | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | Y / N |
| Opiate/Narcotic | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | Y / N |
| Surgery (type: _____) | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | Y / N |
| Hospitalization | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | Y / N |
| Other | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | Y / N |

Medical Services Utilization

These questions ask how often your child has been to a health care provider in the past 3 months.

| Did your child make a visit to the health care provider primarily **because of pain in the past 3 months**? | |
|---|---|
| Physician | |
| Nurse practitioner | |
| Physical therapist | |
| Occupational therapist | |
| Counselor/psychologist/psychiatrist | |
| Acupuncturist | |
| Massage therapist | |
| Chiropractor | |
| Naturopath | |
| Emergency room | |
| Hospitalization | |
| Surgeries | |
| Any other provider: | |

Please list the name of all medications taken by your child in the past week (do not include vitamins or stool softeners).

_____

_____

How satisfied are you with the pain treatment your child has received?

❒ Completely satisfied
❒ Very satisfied
❒ Satisfied
❒ Dissatisfied
❒ Very dissatisfied
❒ Completely dissatisfied

Figure 10: Treatment History Questionnaire

*Data Analysis*

Kinematic Data Analysis

Nine participants were utilized for kinematic data analysis. The first process of kinematic data analysis was completed using Vicon Nexus. The markers were labeled in Vicon based on the anatomical marker set (Figure 5). Throughout the tasks, there are times in which the movement of the participant covers the retroreflective markers. When this happens, the cameras are unable to see the marker, causing a gap in the kinematic data. Within Vicon, all gaps of less than twenty frames were filled using an appropriate mathematical equation within the software. Once all markers were labeled and all gaps were filled, the trials were processed through a Woltring Filter (Schnorenberg et al., 2014). The next component of data analysis was identifying the start and end frames of each trial for every subject. This was completed through visualization of the motion capture trial; the start frame was determined based on when the participant began the movement of the task. The end frame was then determined when the participant returned

their hands to their sides. This process is based on previous studies of joint kinematics (Klotz et al., 2013; Mackey, Walt, Lobb, & Stott, 2005; van Andel, Wolterbeek, Doorenbosch, Veeger, & Harlaar, 2008). Three trials of each task were then selected for each participant for further analysis. Trials were selected based on minimal or absent marker gaps and movement quality. A biomechanical model for pediatric upper extremity joint kinematics was utilized to identify joint angles at the wrist, glenohumeral joint, and thoracohumeral joint throughout each task (Schnorenberg et al., 2014). This model was processed using MATLAB (Mathworks, Inc., Natick, MA) and follows ISB recommendations for rotation sequences (Wu et al., 2005). The model utilized a Y-X-Z Euler sequence for the upper extremity joints (Table 6). Once data were modeled, the peak joint angles and range of motion for each task at the wrist joint, glenohumeral joint, and thoracohumeral joint were calculated in Excel. Subject and group averages and standard deviations were then calculated in Excel.

Table 7: Joint Axes of the Wrist Joint, Glenohumeral Joint, and Thoracohumeral Joint

| Glenohumeral Joint Axes | | Wrist Joint Axes | | Thoracohumeral Joint Axes | |
|---|---|---|---|---|---|
| +X | Adduction | +X | Ulnar Deviation | +X | Scapular Upward Rotation |
| -X | Abduction | -X | Radial Deviation | -X | Scapular Downward Rotation |
| +Y | Internal Rotation | +Y | Internal Rotation (Pronation) | +Y | Scapular Internal Rotation (Protraction) |
| -Y | External Rotation | -Y | External Rotation (Supination) | -Y | Scapular External Rotation (Retraction) |
| +Z | Flexion | +Z | Flexion | +Z | Posterior Spinal Tilt |
| -Z | Extension | -Z | Extension | -Z | Anterior Spinal Tilt |

Pain Location and Severity Questionnaire Data Analysis

All eleven participants were utilized for data analysis of the Pain Location and Severity Questionnaire. The data collected was digitized into and Excel spreadsheet. Any identifying information was omitted from the spreadsheet. Data regarding the participant's and their legal

guardian's answers to each joint and bodily location related to their pain severity. The data was then organized to identify the quantity of bodily locations that the participant identified as a location of pain and the NRS value of their maximal report of pain. Maximal reports of pain that were greater than 4/10 were then counted and reported out of the entire participant group. A score above 4/10 on the NRS considered a moderate to severe pain level, identifying a level of pain in which a treatment intervention is often necessary (Miró et al., 2009)

Treatment History Questionnaire

All eleven legal guardians of the children with hEDS were utilized for data analysis of the Treatment History Questionnaire. All data were digitized and organized into an Excel spreadsheet. Any identifying information was omitted from the spreadsheet. All information regarding the treatments, treatment frequency, and level of satisfaction was recorded and organized. The quantity of treatments was counted and organized per participant. Further, each answer to level of satisfaction was counted and the mode answer was identified using Excel functions.

Functional Disability Inventory Data Analysis

All eleven participants were utilized for data analysis of the Functional Disability Inventory. The Functional Disability Inventory was scored based on author instructions (Kashikar-Zuck et al., 2011). The scores were then organized within an Excel spreadsheet with personal information omitted from all data entered. Scores interpretation was then completed based on previous research. A score of less than 12 identified minimal perceived disability, scores 13-29 identified moderate perceived disability, and any score greater than 30 identified severe perceived disability due to pain (Flowers & Kashikar-Zuck, 2011). To identify a relationship between NRS score of each child's report of worst pain and the FDI scores, a

Pearson product moment correlation test was completed in Excel. The Pearson product moment correlation evaluates the relationship between two variables. A strong relationship will yield an r value of .50 to .75 or above. Alternatively, a weaker relationship will yield an r value less than .50.

## Results

### *Kinematic Data*

The kinematic data presented is from the wrist, glenohumeral, and thoracohumeral joints. The glenohumeral and thoracohumeral joints both depict shoulder movement from two separate perspectives. The glenohumeral joint describes the humerus motion relative to the scapula. Analyzing the shoulder from this perspective can be beneficial when comparing to previous literature on kinematic data, however, it may not represent the more clinical presentation of joint movements. The thoracohumeral joint describes the movement of the shoulder as the humerus relative to the thorax. This measurement offers values that would be similar to goniometric measurements, which may, in turn be more applicable to clinical practice. However, data from the thoracohumeral perspective may be slightly inaccurate of true peak values as the placement of the markers on the body offset the starting position of the individual. This can in turn make the values display the participants' motion as more forward. Regardless, the wrist and shoulder joints are especially significant in this population, as these joints are often cited as common locations for pain and hypermobility (Engelbert et al., 2017; Malfait et al., 2017)

#### Drinking Task

During the drinking task, the wrist was primarily in radial deviation, reaching about 11° (± 8.4°), and in extension, reaching approximately 56° (±4.9°). The glenohumeral joint displayed the most movement within the transverse plane, reaching approximately 65° (± 17.1°) of external

rotation. Data from the thoracohumeral joint displayed its largest peak during flexion, reaching 80° (± 31°) with external rotation being the second largest with 36° (±16.4°) (Figure 10, Table 8).

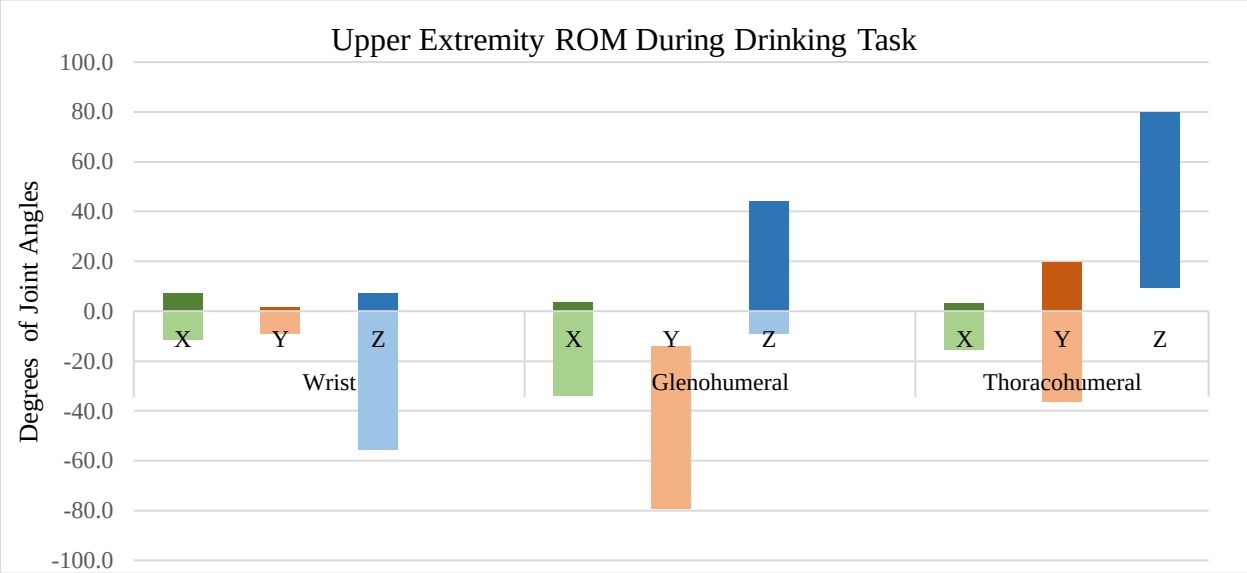



Figure 11: Upper Extremity Range of Motion During Drinking Task

*X=Coronal Angle, Y=Transverse Angle, Z=Sagittal Angle
Mean values for the group maximal, minimal, and range of motion joint values were provided. At the wrist coronal angle positive/negative values are ulnar/radial deviation, transverse angle positive/negative values are pronation/supination, and sagittal angle positive/negative are flexion/extension. At the glenohumeral joint coronal angle positive/negative values are adduction/abduction, transverse angle positive/negative values are internal/external rotation, and sagittal angle positive/negative values are flexion/extension. At the thoracohumeral joint coronal angle positive/negative values are adduction/abduction, , transverse angle positive/negative values are internal/external rotation, and sagittal angle positive/negative values are flexion/extension.

Table 8: Upper Extremity Kinematics During Drinking Task

| Upper Extremity Kinematics During Drinking Task | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|
| | Wrist | | | Glenohumeral Joint | | | Thoracohumeral Joint | | |
| | CA* | TA* | SA* | CA* | TA* | SA* | CA* | TA* | SA* |
| Avg Group Max | 7.1 ± 8.4 | 1.7 ± 2.0 | 7.0 ± 13.6 | 3.6 ± 7.4 | -14.2 ± 11.3 | 44.3 ± 19.8 | 3.2 ± 4.2 | 19.5 ± 11.0 | 80.0 ± 31.0 |
| Avg Group Min | -11.3 ± 6.2 | -8.9 ± 3.9 | -55.7 ± 4.9 | -33.9 ± 7.9 | -65.2 ± 17.1 | -9.1 ± 11.6 | -15.5 ± 7.1 | -36.2 ± 16.4 | 9.4 ± 14.7 |

| | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|
| Avg Group ROM | 19.2 ± 4.2 | 10.9 ± 2.8 | 63.5 ± 12.2 | 37.5 ± 10.0 | 50.9 ± 15.2 | 53.3 ± 12.9 | 18.7 ± 4.8 | 55.8 ± 18.2 | 70.6 ± 10.0 |

*CA=Coronal Angle, TA = Transverse Angle, SA=Sagittal Angle

All values listed are represent the degree of the average group joint angle and the maximum, minimum, range of motion and standard deviations during the task. At the wrist coronal angle positive/negative values are ulnar/radial deviation, transverse angle positive/negative values are pronation/supination, and sagittal angle positive/negative are flexion/extension. At the glenohumeral joint coronal angle positive/negative values are adduction/abduction, transverse angle positive/negative values are internal/external rotation, and sagittal angle positive/negative values are flexion/extension. At the thoracohumeral joint coronal angle positive/negative values are adduction/abduction, transverse angle positive/negative values are internal/external rotation, and sagittal angle positive/negative values are flexion/extension

Combing Task

Data analysis at the thoracohumeral joint for one participant was excluded due to modeling errors. During the combing task, the primary movements were at the glenohumeral and thoracohumeral joints. At the glenohumeral joint, shoulder abduction reached 64° (±15.0), external rotation reached 94° (±32.5°), and maximal shoulder flexion was 52° (±48.9°). From the perspective of the thoracohumeral joint, the participants were primarily in shoulder flexion. Maximum flexion was greater at the thoracohumeral joint, reaching 142° (±37.3°). At the wrist, ulnar deviation reached an average maximal value of 20° (±9.8°) while extension reached 37° (±16.2). (Figure 11, Table 9).

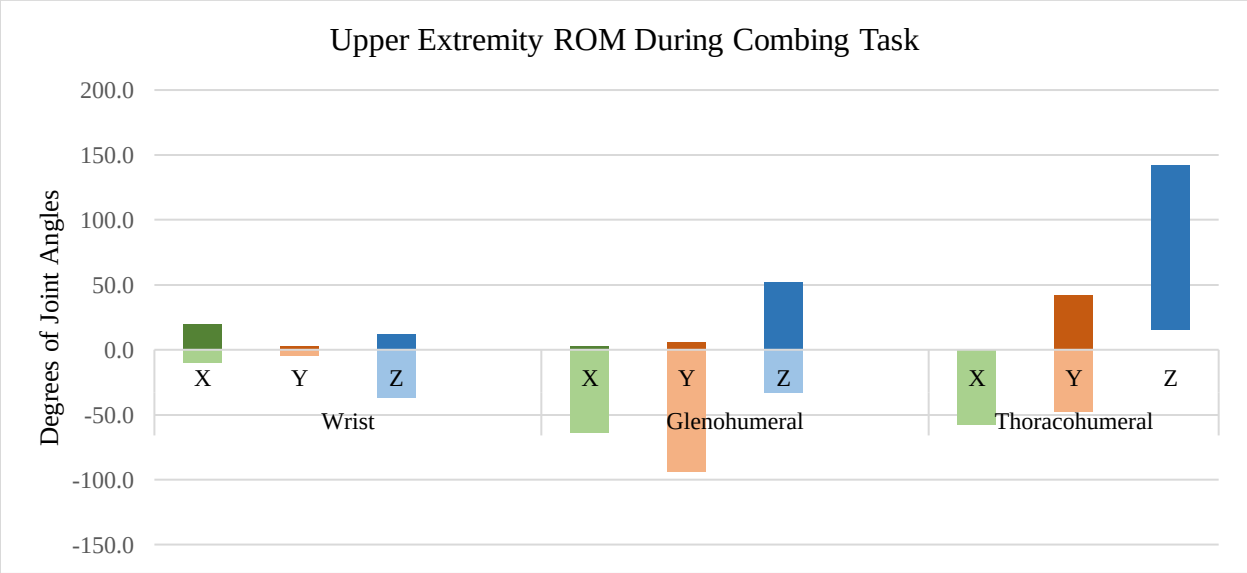


Figure 12: Upper Extremity Range of Motion During Combing Task

*X=Coronal Angle, Y=Transverse Angle, Z=Sagittal Angle
Mean values for the group maximal, minimal, and range of motion joint values were provided. At the wrist coronal angle positive/negative values are ulnar/radial deviation, transverse angle positive/negative values are pronation/supination, and sagittal angle positive/negative are flexion/extension. At the glenohumeral joint coronal angle positive/negative values are adduction/abduction, transverse angle positive/negative values are internal/external rotation, and sagittal angle positive/negative values are flexion/extension. At the thoracohumeral joint coronal angle positive/negative values are adduction/abduction, , transverse angle positive/negative values are internal/external rotation, and sagittal angle positive/negative values are flexion/extension.

Table 9: Upper Extremity Kinematics During Combing Task

| Upper Extremity Kinematics During Combing Task | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|
| | Wrist | | | Glenohumeral | | | Thoracohumeral | | |
| | CA | TA | SA | CA | TA | SA | CA | TA | SA |
| Avg Group Max | 19.7± 9.8 | 2.9 ± 2.3 | 11.9 ± 19.7 | 3.0 ± 7.0 | 6.0 ± 49.8 | 51.9 ± 48.9 | -0.5 ± 7.6 | 42.1 ± 25.5 | 141.6 ± 37.3 |
| Avg Group Min | -9.8 ± 7.4 | -4.7 ± 3.1 | -37.1 ± 16.2 | -63.6 ± 15.0 | -94.3 ± 32.5 | -33.4 ± 40.7 | -57.4 ± 19.3 | -48.0 ± 24.5 | 15.4 ± 11.8 |
| Avg Group ROM | 29.5 ± 5.3 | 7.6 ± 3.6 | 49.0 ± 25.7 | 66.7 ± 13.8 | 100.3 ± 65.8 | 85.3 ± 75.4 | 56.9 ± 23.7 | 90.1 ± 31.8 | 126.3 ± 33.1 |

*CA=Coronal Angle, TA = Transverse Angle, SA=Sagittal Angle
All values listed are represent the degree of the average group joint angle and the maximum, minimum, range of motion and standard deviations during the task. At the wrist coronal angle positive/negative values are ulnar/radial deviation, transverse angle positive/negative values are pronation/supination, and sagittal angle positive/negative are flexion/extension. At the glenohumeral joint coronal angle positive/negative values are adduction/abduction, transverse angle positive/negative values are internal/external rotation, and sagittal angle positive/negative values are flexion/extension. At the thoracohumeral joint coronal angle positive/negative values are adduction/abduction, transverse angle positive/negative values are internal/external rotation, and sagittal angle positive/negative values are flexion/extension

Reaching Across Midline Task

Data from three participants were excluded from analysis of the reaching across midline task. During this task, the trials from these participants had large gaps due to the retro-reflective markers being covered. Throughout the reaching across the midline task, greater ranges of motion were seen at the glenohumeral and thoracohumeral joints. Within the glenohumeral joint maximal external rotation of 66° (±13.9°) was greater than average maximal internal rotation of 42° (±8.9°). During the task, the participants did not engage in glenohumeral extension,

reaching 55° (± °) of average maximal flexion. At the thoracohumeral joint, internal rotation reached 81° (± 8.6) while shoulder flexion reached 76° (±8.3)(Figure 12, Table 10).

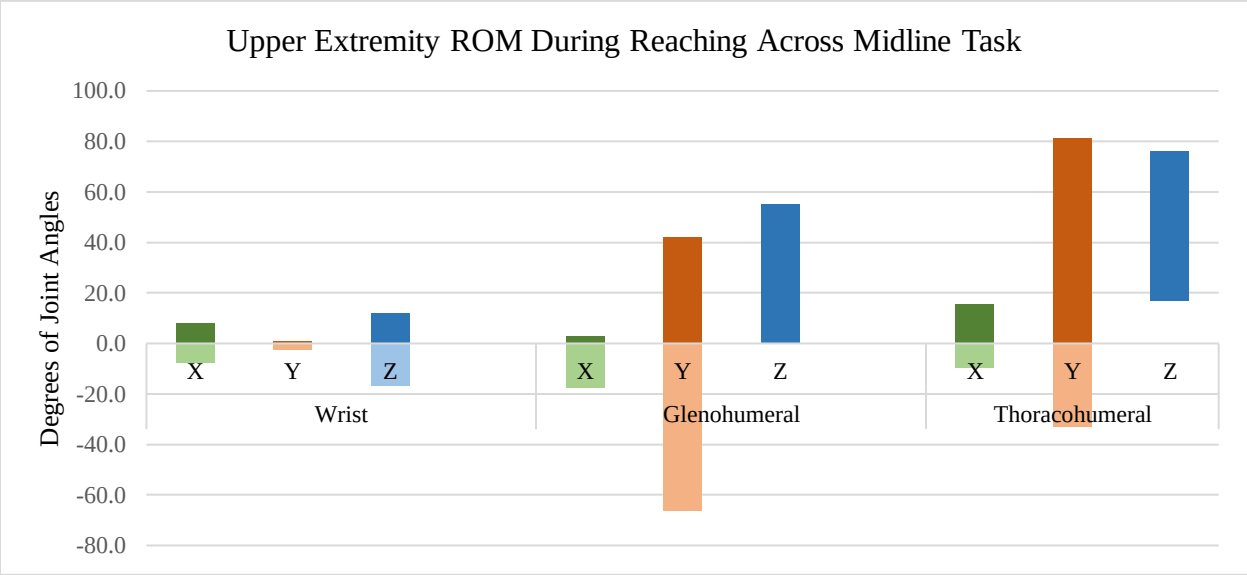



Figure 13: Upper Extremity Range of Motion During Reaching Across Midline Task

*X=Coronal Angle, Y=Transverse Angle, Z=Sagittal Angle
Mean values for the group maximal, minimal, and range of motion joint values were provided. At the wrist coronal angle positive/negative values are ulnar/radial deviation, transverse angle positive/negative values are pronation/supination, and sagittal angle positive/negative are flexion/extension. At the glenohumeral joint coronal angle positive/negative values are adduction/abduction, transverse angle positive/negative values are internal/external rotation, and sagittal angle positive/negative values are flexion/extension. At the thoracohumeral joint coronal angle positive/negative values are adduction/abduction, , transverse angle positive/negative values are internal/external rotation, and sagittal angle positive/negative values are flexion/extension.

Table 10: Upper Extremity Kinematics During Reaching Across Task

| Upper Extremity Kinematics During Reaching Across Task | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|
| | Wrist | | | Glenohumeral | | | Thoracohumeral | | |
| | CA | TA | SA | CA | TA | SA | CA | TA | SA |
| Avg Group Max | 8.1 ± 5.3 | 1.0 ± 0.8 | 12.2 ± 12.4 | 2.9 ± 5.6 | 42.2 ± 8.9 | 54.7 ± 9.1 | 15.7 ± 2.8 | 81.4 ± 8.6 | 76.0 ± 8.3 |
| Avg Group Min | -7.6 ± 4.5 | -2.5 ± 1.3 | -17.0 ± 14.2 | -17.6 ± 5.2 | -66.3 ± 13.9 | 0.2 ± 6.0 | -9.9 ± 6.9 | -33.3 ± 10.9 | 16.7 ± 6.5 |
| Avg Group ROM | 15.8 ± 6.1 | 3.5 ± 2.2 | 29.2 ± 19.1 | 20.6 ± 7.2 | 108.5 ± 44.3 | 54.6 ± 25.3 | 25.5 ± 12.7 | 114.6 ± 46.7 | 59.4 ± 27.6 |

*CA=Coronal Angle, TA = Transverse Angle, SA=Sagittal Angle

All values listed are represent the degree of the average group joint angle and the maximum, minimum, range of motion and standard deviations during the task. At the wrist coronal angle positive/negative values are ulnar/radial deviation, transverse angle positive/negative values are pronation/supination, and sagittal angle positive/negative are flexion/extension. At the glenohumeral joint coronal angle positive/negative values are adduction/abduction, transverse angle positive/negative values are internal/external rotation, and sagittal angle positive/negative values are flexion/extension. At the thoracohumeral joint coronal angle positive/negative values are adduction/abduction, transverse angle positive/negative values are internal/external rotation, and sagittal angle positive/negative values are flexion/extension

## Reaching Back to Ipsilateral Side Task

During the reaching back task, the average maximal wrist ulnar deviation reached 19° (±9.2°) and the average maximal wrist extension reached 42° (±16.6°). At the glenohumeral joint average maximal adduction was 20° (±6.3°), with little abduction of 6° (± 10.5°) identified during this task. Glenohumeral external rotation reached 48° (± 14.4°) and average maximal extension was 46° ± °). At the thoracohumeral joint, the average maximum shoulder abduction was 24°(±16.2), while extension reached 23°. Further, internal and external rotation reached 37° (± 16.6°) and 22° (±13.6) respectively.

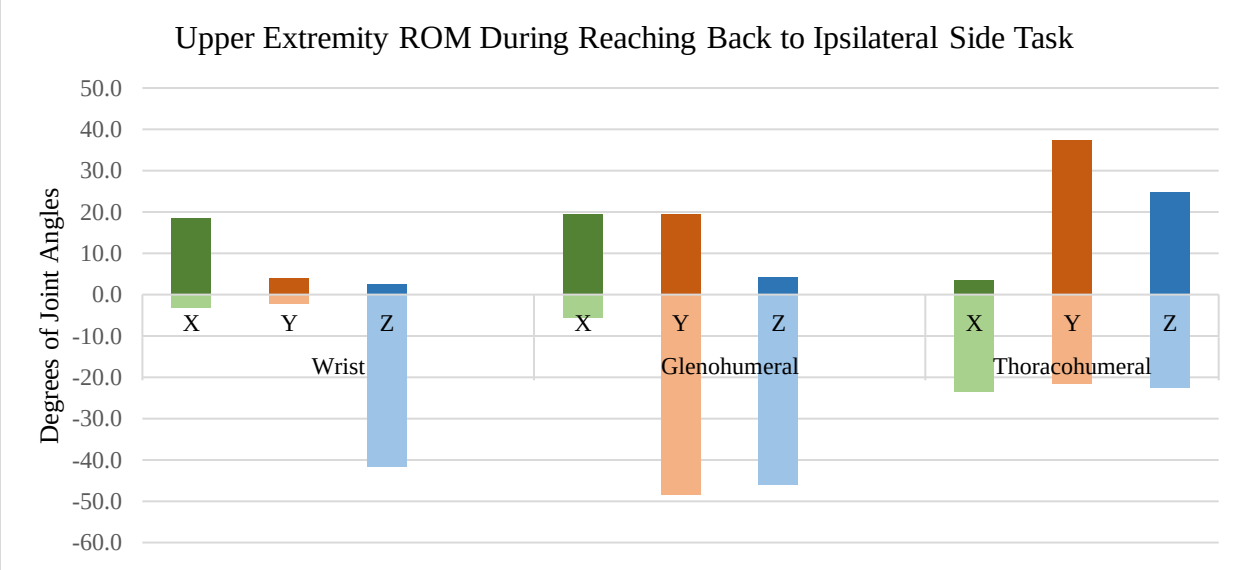



Figure 14: Upper Extremity Range of Motion During Reaching Back to Ipsilateral Side Task

*X=Coronal Angle, Y=Transverse Angle, Z=Sagittal Angle
Mean values for the group maximal, minimal, and range of motion joint values were provided. At the wrist coronal angle positive/negative values are ulnar/radial deviation, transverse angle positive/negative values are pronation/supination, and sagittal angle positive/negative are flexion/extension. At the glenohumeral joint coronal

angle positive/negative values are adduction/abduction, transverse angle positive/negative values are internal/external rotation, and sagittal angle positive/negative values are flexion/extension. At the thoracohumeral joint coronal angle positive/negative values are adduction/abduction, , transverse angle positive/negative values are internal/external rotation, and sagittal angle positive/negative values are flexion/extension.

Table 11: Upper Extremity Kinematics During Reaching Back Task

| Upper Extremity Kinematics During Reaching Back Task | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|
| | Wrist | | | Glenohumeral | | | Thoracohumeral | | |
| | CA | TA | SA | CA | TA | SA | CA | TA | SA |
| Avg Group Max | 18.6 ± 9.2 | 4.1 ± 3.0 | 2.6 ± 7.0 | 19.5 ± 6.3 | 19.4 ± 8.1 | 4.3 ± 10.4 | 3.5 ± 7.6 | 37.4 ± 16.6 | 24.7 ± 13.1 |
| Avg Group Min | -3.1 ± 4.9 | -2.3 ± 1.8 | -41.8 ± 16.6 | -5.7 ± 10.5 | -48.4 ± 14.4 | -46.1 ± 7.9 | -23.5 ± 16.2 | -21.5 ± 13.6 | -22.5 ± 9.9 |
| Avg Group ROM | 21.7 ± 5.5 | 6.4 ± 2.8 | 44.3 ± 15.0 | 25.2 ± 7.6 | 67.7 ± 18.5 | 50.4 ± 11.3 | 27.0 ± 13.2 | 58.9 ± 20.4 | 47.2 ± 9.4 |

*CA=Coronal Angle, TA = Transverse Angle, SA=Sagittal Angle
All values listed are represent the degree of the average group joint angle and the maximum, minimum, range of motion and standard deviations during the task. At the wrist coronal angle positive/negative values are ulnar/radial deviation, transverse angle positive/negative values are pronation/supination, and sagittal angle positive/negative are flexion/extension. At the glenohumeral joint coronal angle positive/negative values are adduction/abduction, transverse angle positive/negative values are internal/external rotation, and sagittal angle positive/negative values are flexion/extension. At the thoracohumeral joint coronal angle positive/negative values are adduction/abduction, transverse angle positive/negative values are internal/external rotation, and sagittal angle positive/negative values are flexion/extension

**Aim 1 Hypothesis: It is hypothesized that during the ADL, children with hEDS will display greater glenohumeral, wrist, and thoracohumeral range of motion when compared to existing kinematic literature.**

Summary of Aim 1 Results

Results from the kinematic data identify that at the glenohumeral joint, flexion was greatest during the reach across task, while abduction and external rotation were reached greater maximal angles during the combing task. Minimal internal rotation was needed during the four tasks, in which the drinking task did not utilize glenohumeral internal rotation. At the wrist joint, the highest maximal angles for flexion and supination were during the drinking task, and the greatest ulnar deviation was during the combing task. At the thoracohumeral joint, maximal flexion, abduction, internal rotation was seen during the combing task.

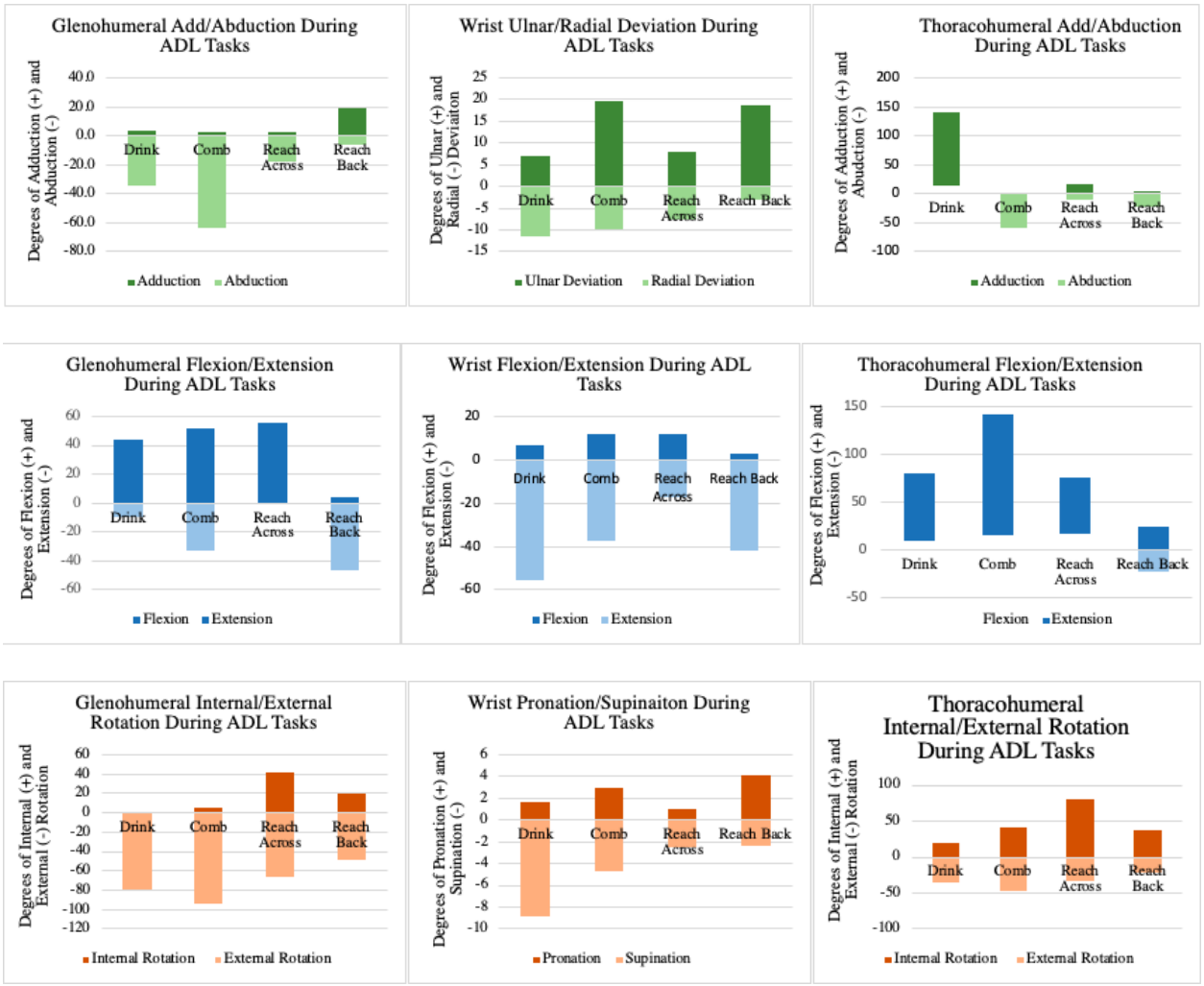



Figure 15: Glenohumeral, Wrist, and Thoracohumeral Range of Motion During ADL Tasks

*X=Coronal Angle, Y=Transverse Angle, Z=Sagittal Angle
Mean values for the group maximal, minimal, and range of motion joint values were provided. At the wrist coronal angle positive/negative values are ulnar/radial deviation, transverse angle positive/negative values are pronation/supination, and sagittal angle positive/negative are flexion/extension. At the glenohumeral joint coronal angle positive/negative values are adduction/abduction, transverse angle positive/negative values are internal/external rotation, and sagittal angle positive/negative values are flexion/extension. At the thoracohumeral joint coronal angle positive/negative values are adduction/abduction, , transverse angle positive/negative values are internal/external rotation, and sagittal angle positive/negative values are flexion/extension.

*Pain Location and Severity Questionnaire*

Results from the Pain Location and Severity Questionnaire identified the joints and bodily regions in which the children in this study most frequently reported pain (Table 12 & 13).

The most common area of pain was the back, all children within this study (11/11) reported pain in the back region. Further back pain was the most common worst location of pain (3/11) among the group. The ankles and throat were both the second most common region of pain (8/11). Only one child identified pain at the face in the groin region. The highest report of worst pain was 10/10 at the region of the head.

Table 12: Participant Reports of Self-Reported Pain Locations

| | 1 | 2 | 3 | 4 | 5 | 6 | 7 | 8 | 9 | 10 | 11 | Total |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| Shoulder | X | X | X | X | | | X | | | | | 5 |
| Wrist | X | X | X | X | | X | X | | | | | 6 |
| Neck | | | X | | | X | X | X | X | X | | 6 |
| Jaw | X | | X | X | | X | | | X | X | X | 7 |
| Elbow | X | | X | X | | X | | | X | | | 5 |
| Fingers | X | | X | X | | X | X | | X | X | | 7 |
| Hips | X | X | X | | | X | X | | X | | | 6 |
| Knees | X | | X | X | | X | X | | | X | X | 7 |
| Ankles | X | X | X | X | | X | X | X | X | | | 8 |
| Toes | | | X | | | X | | | | | | 2 |
| Head | | X | X | X | X | X | X | | X | | | 7 |
| Face | | | X | | | | | | | | | 1 |
| Throat | X | X | X | | X | X | X | X | X | | | 8 |
| Chest | | X | X | X | X | X | X | X | | | | 7 |
| Arms | X | | X | X | | X | X | X | | X | | 7 |
| Hand | X | | X | | | X | X | | | X | | 5 |
| Abdomen | X | X | X | | | X | X | X | X | | | 7 |
| Groin | X | | | | | | | | | | | 1 |
| Legs | X | | X | X | | X | X | | | X | | 6 |
| Feet | | | X | | | X | X | | | | | 3 |
| Back | X | X | X | X | X | X | X | X | X | X | X | 11 |
| Total | 15 | 9 | 20 | 12 | 4 | 18 | 16 | 7 | 10 | 8 | 3 | |
| Average | 11 ± 5 | | | | | | | | | | | |

An ‘X’ identifies whether the child answered ‘Yes’ to whether they experienced pain at that specified location. The average amount of pain locations was 11 ± 5.

Table 13: NRS Self-Reported Pain Scores

| | 1 | 2 | 3 | 4 | 5 | 6 | 7 | 8 | 9 | 10 | 11 |
|---|---|---|---|---|---|---|---|---|---|---|---|
| Worst Pain Location | Hips | Back | Back | Head | Head | Ankles | Neck | Ankles | Back | Knees | Jaw |
| NRS | 7/10 | 7/10 | 9/10 | 10/10 | 4/10 | 8/10 | 4/10 | 9/10 | 9/10 | 9/10 | 7/10 |

| Score Interpretation | Severe | Severe | Severe | Severe | Mod | Severe | Mod | Severe | Severe | Sever | Severe |
|---|---|---|---|---|---|---|---|---|---|---|---|
| Average | 7.5 ± 2 | | | | | | | | | | |

The location of worst pain was identified based on the location with the highest NRS scores the child provided. Score interpretation was based off of previous studies Mild=mild pain, Mod=moderate pain, Severe=Severe pain.

**Aim 2 Hypotheses A and B: It is hypothesized that children with hEDS will identify multiple locations of pain. It is hypothesized that their highest level of self-reported pain will be greater than 4/10 on the Numeric Rating Scale (NRS).**

Summary of Aim 2 Results

Results of the Pain Location and Severity Questionnaire identified that all children (11/11) within this study reported pain in multiple joints and bodily locations. On average, the participants reported 11 locations of pain (±5 locations). The maximal amount of locations was 20 out of 21 possible bodily locations. Further, results from the NRS identify that all children in this study reported their maximal pain to be greater than 4/10. The average report of maximal pain was 7.5/10 (±2) and all the participants were within the moderate to severe pain ranges. The majority (9/11) of the participants were within the severe pain range. Previous studies identify that pain greater than a 4/10 designates the need for intervention or treatment (Breivik et al., 2008).

*Treatment History Questionnaire*

Results from the Treatment History Questionnaire identify that the average amount of trialed pain treatments for each child was 5 (±3). The maximal amount of treatments trialed was 10/14 different treatments. The most common treatments utilized within the group were exercise (8/11) and ice (7/11). Only one parent reported not using any treatments for pain management. Parents that selected other were asked whether they were comfortable sharing the pain treatment. One parent reported utilizing medical marijuana for the treatment of their child's pain. The mode

answer to the Likert satisfaction scale was 'Satisfied'. No parents reported being completely satisfied or dissatisfied with the treatments that their child had received.

Table 14: Parent Reports for Treatment History Questionnaire

| | 1 | 2 | 3 | 4 | 5 | 6 | 7 | 8 | 9 | 10 | 11 | Total |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| Exercise | X | X | | X | X | X | | X | | X | X | 8 |
| Massage | X | X | | | X | | | | | X | | 4 |
| Heat | X | | | | X | X | | X | X | X | | 6 |
| Ice | X | X | | X | X | X | | X | | X | | 7 |
| TENS | X | | | | | | | | | X | | 2 |
| Relaxation | X | X | | | | | | | | | X | 3 |
| Psychological Counseling | X | X | | X | X | X | | | | | | 5 |
| Antidepressants | | X | X | X | | X | | | | | | 4 |
| Anti-inflammatory | X | X | | | | X | | | X | X | X | 6 |
| Acetaminophen | | | | | | X | | | X | X | | 3 |
| Opiate | | | | | | X | | | | | | 1 |
| Surgery | X | X | | X | | X | | | | | | 4 |
| Hospital | | | | X | | | | | | | | 1 |
| Other | | X | | | | X | | | | X | | 3 |
| Total | 9 | 9 | 1 | 6 | 5 | 10 | 0 | 3 | 3 | 8 | 3 | |
| Average | 5 (± 3) | | | | | | | | | | | |

Table 15: Parent Reports for Treatment Satisfaction

| | 1 | 2 | 3 | 4 | 5 | 6* | 7 | 8 | 9 | 10 | 11 | Total |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| CS | | | | | | | | | | | | 0 |
| VS | | | | | | X | | X | | | | 2 |
| S | X | X | | X | | | | | X | | | 4 |
| DS | | | X | | | | | | | X | X | 3 |
| VD | | | | | X | | | | | | | 1 |
| CD | | | | | | | | | | | | 0 |

*CS=Completely Satisfied, VS=Very Satisfied, S=Satisfied, D=Dissatisfied, VD=Very Dissatisfied, CD =Completely Dissatisfied

**Aim 3 Hypothesis A and B: It is hypothesized each parent will report trialing multiple treatments to manage their child's hEDS pain. It is hypothesized that the average level of satisfaction will be "Dissatisfied" on a Likert satisfaction scale.**

Summary of Aim 3 Results

Results from this study identified that almost all parents trialed multiple forms of treatments (10/11). However, the parent that did not trial any treatments was the parent of the child with the lowest maximal score for pain (4/10). The mode answer to the Likert satisfaction scale was 'Satisfied' (4/10). The parent that reported not trialing any pain treatments was left out of the Likert satisfaction scale.

*Functional Disability Inventory*

Results from the FDI display the level of perceived disability due to pain that the child has regarding themself. Scores were not recorded for participant 1, for comparison to the NRS, a score of 0 was provided to participant 1. The majority of the participants (5/10) identified to having moderate perceived disability due to pain (Table 16 &17). Only one child was within the severe disability range. The lowest score from the FDI was a 1/60, identifying minimal perceived disability.

Table 16: FDI Raw Scores and Score Interpretations

| | 1* | 2 | 3 | 4 | 5 | 6 | 7 | 8 | 9 | 10 | 11 |
|---|---|---|---|---|---|---|---|---|---|---|---|
| FDI Score** | X | 28 | 23 | 23 | 24 | 34 | 3 | 1 | 23 | 9 | 6 |
| Score Interpretation*** | X | Mod | Mod | Mod | Mod | Severe | Min | Min | Mod | Min | Min |

* FDI score for participant 1 was unavailable
**Functional Disability Scores were scored based on author recommendations; a max score on the FDI is 60.
*** Score interpretations were identified based on author recommendations: Min=minimal perceived disability, Mod=moderate perceived disability, Severe=severe perceived disability.

Table 17: FDI and NRS Scores

| | 1 | 2 | 3 | 4 | 5 | 6 | 7 | 8 | 9 | 10 | 11 |
|---|---|---|---|---|---|---|---|---|---|---|---|
| FDI Score | 0* | 28 | 23 | 23 | 24 | 34 | 3 | 1 | 23 | 9 | 6 |
| NRS** | 7/10 | 7/10 | 9/10 | 10/10 | 4/10 | 8/10 | 4/10 | 9/10 | 9/10 | 9/10 | 7/10 |

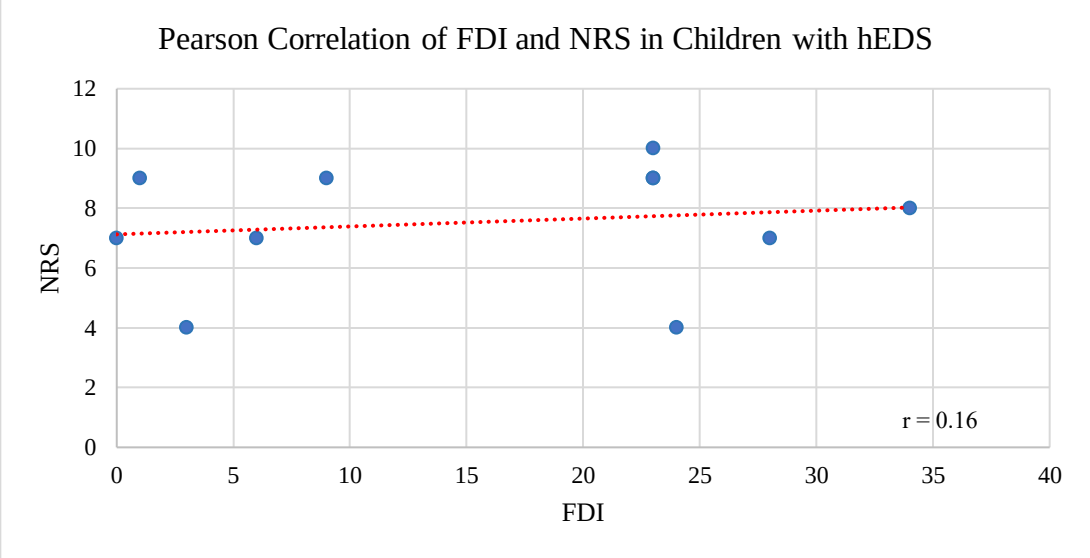
*To perform statistical analyses between the FDI score of participant 1 was designated as 0/60
**The NRS score from each participant's maximal location of pain was utilized.

**Aim 4 Hypothesis: It is hypothesized that children with higher ratings of pain will have higher scores of perceived disability in the Functional disability inventory**

Summary of Aim 4 Results

A Pearson product moment correlation was conducted to identify a relationship between FDI scores and NRS scores (Figure 15). The correlation coefficient for the FDI and NRS resulted in r=0.16, identifying a weak relationship between the two variables. Further, the coefficient of determination $r^2$=0.025 identified that only 2.6% of the information from the FDI can assist in determining the NRS scores. The p-value was found to be 0.64, whereas, a p-value of p<0.05 would be appropriate. This p-value identifies that the small sample size used in this study limits its ability to generalize to a larger population (Portney & Watkins, 2015).

Figure 16: Pearson Correlation of FDI and NRS




*P value=0.64, $r^2$=0.025

## Discussion

*Kinematics*

Collecting normative pediatric upper extremity data for comparison purposes in this study was paused due to IRB restrictions for human subject testing due to Coronavirus Disease 2019 (COVID-19). For the purposes of this study, existing upper extremity kinematic literature in both pediatric and adult populations were utilized to make qualitative comparisons of upper extremity kinematics during ADL tasks (Table 18) (Gates, Walters, Cowley, Wilken, & Resnik, 2016; Mackey, Walt, & Stott, 2006; Petuskey, Bagley, Abdala, James, & Rab, 2007; van Andel et al., 2008). The studies identified reported their kinematic data at a variety of joints during a variety of upper extremity tasks. Further, each study utilized a different motion capture system and retroreflective marker set for their data collection. Additionally, some studies reported on shoulder motion from the glenohumeral joint, while others reported from the thoracohumeral joint. Because of the variability between the studies, formal statistical comparisons were not feasible and therefore were not completed. Rather, the identification of clinically significant differences and noteworthy findings are reported.

Table 18: Upper Extremity Kinematic Research

| | Age Group | N | Tasks | Joint Reported | Start/End Positions | Motion Capture |
|---|---|---|---|---|---|---|
| Mackey 2006 | Pediatric | 10 | Hand-to-mouth<br>Hand-to-Head<br>Reaching Task | Elbow Flexion/Extension<br>Elbow Rotation<br>Shoulder Flexion/Extension<br>Shoulder Ab/Adduction<br>Trunk Flexion/Extension | Start and end with hands positioned on a table | 8 camera system<br>21 retroreflective markers<br>60Hz |
| Petuskey 2007 | Pediatric | 51 | Hand to Back Pocket<br>Hand to Top of Head<br>High Reach<br>Forward Reach<br>Wave (arm at side shoulder ext. rotated) | Shoulder Flexion/Extension, Ab/Adduction, Int/Ext Rotation<br>Elbow Flexion/Extension, Pronation/Supination<br>Neck Rotation, Flexion | Start and end positioned with arms relaxed at sides | 8 camera system<br>18 retroreflective markers<br>60Hz |

| | | | | | | |
|---|---|---|---|---|---|---|
| Van Andel 2008 | Adults | 10 | Drinking<br>Hand to Back Pocket<br>Hand to Contralateral Shoulder<br>Combing | Scapular Protraction/Retraction, Lateroroation, Tilt<br>Humeral Elevation Plane, Elevation, Rotation<br>Elbow Flexion/Extension, Pronation/Supination<br>Wrist Flexion/Extension, Radioulnar Deviation<br>Trunk Flexion, Lateral Flexion, Axial Rotation | Start with hands relaxed at sides (except for drinking task, the hand was holding a cup relaxed on knee) | 3 camera system<br>19 LED markers<br>50Hz |
| Gates 2016 | Adults | 15 | Drinking<br>Hand to back pocket<br>Box off Shelf<br>Can off shelf<br>Deodorant<br>Perineal care<br>Box off ground | Humeral Elevation Plane, Elevation, Rotation<br>Elbow Flexion/Extension, Pronation/Supination<br>Wrist Flexion/Extension, Ulnar/Radial Deviation | Authors did not disclose | 38 retroreflective markers<br>120Hz |
| UWM 2020 | Pediatric | 9 | Drinking<br>Reach Back to Ipsilateral Side<br>Combing<br>Reach Across Midline | Scapular Protraction/Retraction, Tilt<br>Shoulder Flexion/Extension, Ab/Adduction, Int/Ext Rotation<br>Wrist Flexion/Extension, Ulnar/Radial Deviation | Start and end with arms positioned at | 15 camera system<br>15 retroreflective markers<br>120Hz |

The kinematic data from the studies were compiled to identify differences in maximal joint angles during specific ADL tasks (Table 19). To establish clinical significance of the kinematic data identified, a difference of 5° was utilized based on previous research (Groth, VanDeven, Philips, & Ehretsman, 2001). During the drinking task, shoulder external rotation displayed clinically significant differences between the hEDS group (65°) when compared to Gates (2016) (53°). Additionally, wrist extension was 56° in the hEDS group, displaying a clinically significant difference when compared to Van Andel (2008) and Gates (2016), 19° and 33° respectively. During the reaching back task, the hEDS group displayed greater peak shoulder extension when compared to Petuskey (2007). Further, wrist extension and scapular protraction were greater in the hEDS group when compared to Gates (2016) and Van Andel (2008). During the drinking task, Mackey (2006) reported greater shoulder flexion, however, later reported the

necessary shoulder flexion to complete the task was 48° which is similar to the findings from this study 44°. Gates (2016) collected data on seven tasks and reported that all tasks could be completed with 38° of wrist flexion, 40° of wrist extension, 38° of ulnar deviation, and 28° of radial deviation. Further, Gates (2016) expresses that all their tasks could be completed with 79° of internal rotation and 55° of external rotation. However, the kinematic data from this study does not always fall within those ranges at the wrist or shoulder.

Throughout the upper extremity ADL tasks, at times the literature displayed greater maximum joint angles when compared to the hEDS. While this information may be contradictory of the claims of hypermobility in children with hEDS, there may also be data collection characteristics that attribute to this finding. For example, Mackey (2006) explains that their shoulder flexion may be greater due to having their participants' start the task with their arm resting on a table. All of the studies mentioned above describe high variability in kinematics throughout their participants (Gates et al., 2016; Mackey et al., 2006; Petuskey et al., 2007; van Andel et al., 2008). The completion of the ADL tasks is not completed in one specific way, resulting in high variability in the maximum and minimum joint angles. Further, each study had their own requirements for the completion of the task. For the drinking task, some studies required the participant to hold a cup or water bottle and pretend to take a full drink. However, other studies only had the participants mimic bringing a small item to their mouth which may limit the necessary shoulder flexion and wrist deviation. Lastly, each study may describe the same task differently to their participants, and each participant may interpret that description in their own way. This component alone may greatly influence joint angles during each task as instructing a participant to reach their hand into their back pocket may require different wrist involvement when compared to instructing a participant to reach back to perform toilet hygiene.

Because of the variability in previous research, it is not appropriate to draw conclusions on whether the children with hEDS from this study have a greater range of motion when compared to a normal population of children or adults. Future studies are warranted to obtain kinematics from a normal population to identify true statistical differences between the children with hEDS and a normal pediatric population. Further, obtaining these data under the same protocol may be beneficial to limit additional variability between data collection. Lastly, a larger sample of both children with hEDS and a group of children without hypermobility may allow for stronger and more reliable results.

Table 19: Comparisons of Upper Extremity Kinematics During ADL Tasks

| Drinking Task | | | | | |
|---|---|---|---|---|---|
| | Mackey 2006 | Van Andel 2008 | Petuskey 2007 | Gates 2016 | UWM 2020 |
| GH Shoulder Flexion | 74° | | | | 44° |
| GH Shoulder Extension | | | | | 9° |
| GH Shoulder Abduction | 41° | | | | 34°* |
| GH Shoulder Adduction | | | | | 4° |
| GH Shoulder Internal Rotation | | | | | 14° |
| GH Shoulder External Rotation | | | | | 65°* |
| TH Shoulder Flexion | | | | | 80° |
| TH Shoulder Extension | | | | | |
| TH Shoulder Abduction | | | | | 16° |
| TH Shoulder Adduction | | | | | 3° |
| TH Shoulder Internal Rotation | | | | | 20° |
| TH Shoulder External Rotation | | | | 53° | 36° |
| Wrist Flexion | | 6.5° | | 8° | 7° |
| Wrist Extension | | 19° | | 33° | 56°* |
| Ulnar Deviation | | 25° | | 23° | 7° |

| Radial Deviation | | | | 11° | 11° |
|---|---|---|---|---|---|
| Reaching Back to Ipsilateral Side | | | | | |
| | Mackey 2006 | Van Andel 2008 | Petuskey 2007 | Gates 2016 | UWM 2020 |
| GH Shoulder Flexion | | | | | 4° |
| GH Shoulder Extension | | | | | 46° |
| GH Shoulder Abduction | | | | | 6° |
| GH Shoulder Adduction | | | | | 20°* |
| GH Shoulder Internal Rotation | | | | 79° | 19° |
| GH Shoulder External Rotation | | | | 53° | 48° |
| TH Shoulder Flexion | | | | | 25° |
| TH Shoulder Extension | | | 47 | | 23° |
| TH Shoulder Abduction | | | 22 | | 24° |
| TH Shoulder Adduction | | | 2 | | 4° |
| TH Shoulder Internal Rotation | | | 30 | | 37° |
| TH Shoulder External Rotation | | | 22 | | 22° |
| Wrist Flexion | | 9° | | 28° | 3° |
| Wrist Extension | | 5° | | 15° | 42°* |
| Ulnar Deviation | | 16° | | 35° | 19° |
| Radial Deviation | | 1° | | 7° | 3° |

*Clinically significant difference between maximum joint angles. (Gates et al., 2016; Mackey et al., 2006; Petuskey et al., 2007; van Andel et al., 2008). Gray areas within this chart depict that the corresponding author did not provide or report this information in their study.

*Pain Location and Severity*

Previous studies related to pain and hEDS have focused on the pain within the adult population. However, information reported in previous studies report that pain in adults with hEDS may have started in their adolescent years (Levy, 2004; Sacheti et al., 1997). Specific information surrounding the pediatric pain experience in children with hEDS is limited. The goal of this study was to obtain data regarding location and severity of pediatric hEDS pain and to

identify any common trends in the group's reports of pain. A major result of this study identified that all children in this study (11/11) self-reported pain related to their hEDS with the maximal report of pain in the group being 10/10 on the NRS. Further, all of the children in this study (11/11) reported pain in more than one region of their body, with the most common region being within at their back. On average the children in this study reported pain in 11 different bodily regions, which is greater than a previous study conducted with adults with hEDS that reported an average of 8 locations (Sacheti et al., 1997). Previous studies have reported that physical activity may be cause exacerbated pain in hEDS, however, this study did not identify the cause of the child's pain, rather the presence of it (Engelbert et al., 2017). Some studies have attributed the pain experience of adults with hEDS to hyperalgesia as a result of a sensitized central nervous system (Castori, 2016; L. Rombaut et al., 2014; M. Scheper et al., 2015). However, the presence of this sensitization is unknown within the pediatric hEDS population. Future studies may benefit from obtaining more specific information on activities that exacerbate pain and would benefit from obtaining a larger pediatric sample.

*Treatment History*

A specific treatment protocol for pain in both pediatric and adult hEDS has not yet been identified. However, many studies report the importance of utilizing a holistic medical team involving general physicians, rheumatologists, physical and occupational therapists, psychologists, and any others that are needed (Gazit et al., 2016; Lies Rombaut et al., 2011). Further, previous studies have reported on the efficacy of specific treatment interventions including pharmaceuticals, rehabilitation protocols, surgeries, psychological support, and more (Castori, 2016; CCHMC, 2014; M. C. Scheper et al., 2013; Smith et al., 2014). However, most do not specifically focus on the pediatric population and whether pain was decreased for long-

term after the implementation of the intervention. The goal of this study was to identify common treatment interventions for pain management in pediatric hEDS and to identify whether parents were satisfied with the treatment that was implemented. On average the parents in this study trialed 5 treatments, with the maximum amount of treatments being 10. Only one parent in this study chose not to utilize interventions for pain management. Some children in this study were prescribed opiate and antidepressant medication to assist in pain management. However, the implementation and long-term effects of these pharmaceuticals in the pediatric population has limited research (Dwyer & Bloch, 2019; Matson et al., 2019). When assessing the level of satisfaction of parents, the mode answer to the Likert satisfaction scale was 'Satisfied' with the treatments that they had trialed. However, one parent disclosed that the only treatment that was effective was medical marijuana and that other more traditional pain management strategies were not effective for managing their child's pain. The results of this study identify the variability in pain management for children with hEDS. Future studies are warranted to administer large scale surveys to parents of children with hEDS and to identify objective efficacy of specific treatment regimens for pain management in hEDS.

*Functional Disability Inventory*

The impact of chronic pain often leads to a lower quality of life and an increase in MF/CFS (Castori, 2016; Sacheti et al., 1997). To identify negative impacts of pediatric chronic pain in the hEDS population, the FDI was administered to each child. The FDI provides insight into the child's understanding of their own physical functioning and their own perception of disability due to pain (Flowers & Kashikar-Zuck, 2011; Kashikar-Zuck et al., 2011). This study administered the FDI to 10 children with hEDS. While all children reported to having moderate to severe pain related to their hEDS, the FDI identified that the children within this study do not

have physical limitations due to their pain. The scores from the FDI reported a range of severity from minimal to severe perceived disability, with the maximum score being 34/60. The results of this study identify that NRS scores do not correlate with a child' level of perceived disability or ability to participate in daily activities. While this result did not support the hypothesis, it displays the resiliency in children with hEDS. The children in this study may be experiencing pain at a severe level, however, they feel that they are still able to do common tasks and activities within their daily life. Future studies may focus on broader aspects of psychological impact due to pain such as quality of life assessments with a larger sample.

**Conclusions**

The primary goal of this study was to obtain both kinematic and qualitative information regarding pediatric hEDS. While at some joints, the children with hEDS had greater peak joint angles, this study cannot appropriately conclude that children with hEDS have a greater range of motion during ADL tasks. However, all children within this study reported pain, and all children in this study reported pain at their back. Further, all children reported maximal pain within the moderate to severe pain category. Treatment interventions for pain management within this study were variable, the maximum amount of interventions trialed was 10 and the lowest was 0. Parents most frequently reported being 'Satisfied' with the pain management treatments that they utilized with their child. The children in this study displayed variable scores on the FDI, identifying that pain may not be a predictor of perceived functional disability due to pain.

# III. CONCLUSION

**Summary of Conclusions**

The results of this study identified the kinematic characteristic of children with hEDS during four functional ADL tasks. Next, this study identified that children with hEDS experience

moderate to severe chronic pain, and pain that is frequently in their back. Results from this study identified that parents utilize a variety of treatment interventions for managing their child's pain. The highest amount of treatment interventions utilized was ten while the lowest was zero. Further, the most frequent response to the Likert satisfaction scale was 'Satisfied' when asked about their satisfaction with treatment interventions for pain. Lastly, this study concluded that a child's highest report of pain does not impact their perceived level of disability. The data from this study identified that the scores of the functional disability inventory to not correlate with scores from the NRS reports of worst location of pain.

**Limitations**

The primary limitations of this study included a small sample size, inability to perform normative data collection, lack of range of motion testing, and modeling difficulties. This study utilized a sample size of 11 subjects, of which only 9 were able to be used for kinematic analysis. The small sample of this study is restrictive in that the information obtained from this study cannot be generalized to a larger population of children with hEDS. Further, the inability to collect normative kinematic upper extremity data greatly limited the ability to identify whether the children with hEDS displayed significantly greater range of motion during the four ADL tasks. Additionally, testing of range of motion with goniometry at the primary upper extremity joints would have been beneficial to identify whether the children in this study reached hyperextension in their normal ranges. Lastly, difficulties with Gimbal lock during the reaching across task further limited the kinematic data utilized in data analysis. While these limitations impacted the ability to further compare and generalize the data collected in this study, the information obtained from this study regarding pediatric hEDS continues to be valuable. This

study can act as a stepping stone for many future research studies surrounding hEDS in the pediatric population.

**Future Directions**

The future of research surrounding pediatric hEDS has many opportunities for new discoveries. Because the amount of known information is limited in pediatric hEDS, all studies can offer valuable and helpful information regarding both the diagnostic and treatment aspects of this condition. As this study concludes, there are many new research questions and needed information to expand and supplement what has already been found.

A primary question that was not answered in this study is whether children with hEDS have greater range of motion during ADL when compared to a normative pediatric control group. Obtaining this information would be best and most accurately completed within the same lab using the same kinematic data collection protocol and analysis models. Adding tasks that utilize the smaller intrinsic hand muscles may be beneficial in the pediatric group as handwriting and drawing are common requirements for participation in school. Further, obtaining the full range of motion using goniometry from both an hEDS group and a normative group can identify whether the children with hEDS do reach hyperextension or greater range of motion within the upper extremity. Lastly, obtaining a large sample of both groups will be beneficial in the ability to generalize the results of the study.

The results from this study identified back pain as a common pain location in the children with hEDS. Future studies may benefit from using a large sample survey to identify common bodily locations and severity of pediatric hEDS pain. Further obtaining information on the specific type of pain that the child experiences can help to better define what is most common

and how to best treat the pain. This information can assist in the phenotypic characteristics of hEDS, further assisting in the diagnostic process.

Previous studies have reported specific physical rehabilitative treatments and protocols for children with hEDS, however, the impact of these treatments on pain and fatigue are still unknown. Future studies would benefit from controlled trials of interventions for pediatric pain management in children with hEDS to best understand the effects of a variety of treatment interventions. Additionally, studies that identify early intervention strategies for the identification or treatment of hypermobility can assist in better understanding of when hEDS symptoms start and how they can be most effectively treated at an early age. Ultimately, hEDS continues to be a condition in which little is known regarding effective management throughout the lifespan. Continued research regarding this condition is warranted to best serve the hEDS community.

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# APPENDIX A: PAIN LOCATION AND SEVERITY QUESTIONNAIRE

Pain Location and Severity

In the last 3 months has your child had pain that bothers him/her in his her ___. If yes, what is the typical pain intensity on a scale of 0-10 where 0 equals no pain and 10 is pain as bad as it can be.

Interviewer, decide which is the worst pain location by 0-10 scale. If some are equal, ask subject which is worst. Code worst (a-)

| Primary Pain Location(s): Joints | | | |
|---|---|---|---|
| | No | Yes | Average/typical intensity on a scale of 0-10 where 0 equals no pain and 10 equals pain as bad as it can be? |
| Neck | | | |
| Jaw | | | |
| Shoulders | | | |
| Elbows | | | |
| Wrists | | | |
| Fingers | | | |
| Hips | | | |
| Knees | | | |
| Ankles | | | |
| Toes | | | |

| Primary Pain Location(s): Body Parts | | | |
|---|---|---|---|
| | No | Yes | Average/typical intensity on a scale of 0-10 where 0 equals no pain and 10 equals pain as bad as it can be? |
| Head | | | |
| Face | | | |
| Throat/Neck | | | |
| Chest | | | |
| Arms | | | |
| Hand | | | |
| Abdomen | | | |
| Groin/Pubic Area | | | |
| Legs | | | |
| Feet | | | |

# APPENDIX B: TREATMENT HISTORY QUESTIONNAIRE

**Treatments**

Have you ever sought treatment for your child's pain?

❒ yes
❒ no

If yes, please continue.

| Has your child had any of the following treatments for pain?<br>❒ yes<br>❒ no | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|
| | Had it? | How long did child receive treatment? | How helpful was it?<br>0 = not at all, 5 = extremely | | | | | | | | Still use it? |
| Exercise | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | | | Y / N |
| Massage | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | | | Y / N |
| Heat | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | | | Y / N |
| Ice | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | | | Y / N |
| TENS | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | | | Y / N |
| Relaxation Training | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | | | Y / N |
| Psychological Counseling | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | | | Y / N |
| Antidepressant (e.g., Elavil) | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | | | Y / N |
| Anti-Inflammatory | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | | | Y / N |
| Tylenol/Acetaminophen | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | | | Y / N |
| Opiate/Narcotic | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | | | Y / N |
| Surgery (type: _____) | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | | | Y / N |
| Hospitalization | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | | | Y / N |
| Other | Y / N | | 0 | 1 | 2 | 3 | 4 | 5 | | | Y / N |

Continued on back

Medical Services Utilization

These questions ask how often your child has been to a health care provider in the past **3 months**.

| Did your child make a visit to the health care provider primarily **because of pain in the past 3 months**? | |
|---|---|
| Physician | |
| Nurse practitioner | |
| Physical therapist | |
| Occupational therapist | |
| Counselor/psychologist/psychiatrist | |
| Acupuncturist | |
| Massage therapist | |
| Chiropractor | |
| Naturopath | |
| Emergency room | |
| Hospitalization | |
| Surgeries | |
| Any other provider: | |

Please list the name of all medications taken by your child in the past week (do not include vitamins or stool softeners).

_____

_____

How satisfied are you with the pain treatment your child has received?

- ❒ Completely satisfied
- ❒ Very satisfied
- ❒ Satisfied
- ❒ Dissatisfied
- ❒ Very dissatisfied
- ❒ Completely dissatisfied

# APPENDIX C: FUNCTIONAL DISABILITY INVENTORY

**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 |

# APPENDIX D: GROUP AVERAGE AND STANDARD DEVIATION KINEMATIC DATA FOR ADL

**DRINK**

| | Wrist | | | Glenohumeral | | | Thoracohumeral | | |
|---|---|---|---|---|---|---|---|---|---|
| | X | Y | Z | X | Y | Z | X | Y | Z |
| Group AVG MAX | 7.1 | 1.7 | 7.0 | 3.6 | -14.2 | 44.3 | 3.2 | 19.5 | 80.0 |
| Group AVG MIN | -11.3 | -8.9 | -55.7 | -33.9 | -65.2 | -9.1 | -15.5 | -36.2 | 9.4 |
| Group AVG ROM | 19.2 | 10.9 | 63.5 | 37.5 | 50.9 | 53.3 | 18.7 | 55.8 | 70.6 |
| GROUP STDEV MAX | 8.4 | 2.0 | 13.6 | 7.4 | 11.3 | 19.8 | 4.2 | 11.0 | 31.0 |
| GROUP STDEV MIN | 6.2 | 3.9 | 4.9 | 7.9 | 17.1 | 11.6 | 7.1 | 16.4 | 14.7 |
| GROUP STDEV DROM | 4.2 | 2.8 | 12.2 | 10.0 | 15.2 | 12.9 | 4.8 | 18.2 | 10.0 |

**COMB**

| | Wrist | | | Glenohumeral | | | Thoracohumeral | | |
|---|---|---|---|---|---|---|---|---|---|
| | X | Y | Z | X | Y | Z | X | Y | Z |
| Group MAX | 19.7 | 2.9 | 11.9 | 3.0 | 6.0 | 51.9 | -0.5 | 42.1 | 141.6 |
| Group MIN | -9.8 | -4.7 | -37.1 | -63.6 | -94.3 | -33.4 | -57.4 | -48.0 | 15.4 |
| Group ROM | 29.5 | 7.6 | 49.0 | 66.7 | 100.3 | 85.3 | 56.9 | 90.1 | 126.3 |
| GROUP STDEV MAX | 9.8 | 2.3 | 19.7 | 7.0 | 49.8 | 48.9 | 7.6 | 25.5 | 37.3 |
| GROUP STDEV MIN | 7.4 | 3.1 | 16.2 | 15.0 | 32.5 | 40.7 | 19.3 | 24.5 | 11.8 |
| GROUP STDEV ROM | 5.3 | 3.6 | 25.7 | 13.8 | 65.8 | 75.4 | 23.7 | 31.8 | 33.1 |

**REACH BACK**

| | Wrist | | | Glenohumeral | | | Thoracohumeral | | |
|---|---|---|---|---|---|---|---|---|---|
| | X | Y | Z | X | Y | Z | X | Y | Z |
| Group MAX | 18.6 | 4.1 | 2.6 | 19.5 | 19.4 | 4.3 | 3.5 | 37.4 | 24.7 |
| Group MIN | -3.1 | -2.3 | -41.8 | -5.7 | -48.4 | -46.1 | -23.5 | -21.5 | -22.5 |
| Group ROM | 21.7 | 6.4 | 44.3 | 25.2 | 67.7 | 50.4 | 27.0 | 58.9 | 47.2 |
| GROUP STDEV MAX | 9.2 | 3.0 | 7.0 | 6.3 | 8.1 | 10.4 | 7.6 | 16.6 | 13.1 |
| GROUP STDEV MIN | 4.9 | 1.8 | 16.6 | 10.5 | 14.4 | 7.9 | 16.2 | 13.6 | 9.9 |
| GROUP STDEV ROM | 5.5 | 2.8 | 15.0 | 7.6 | 18.5 | 11.3 | 13.2 | 20.4 | 9.4 |

**REACH ACROSS**

| | Wrist | | | Glenohumeral | | | Thoracohumeral | | |
|---|---|---|---|---|---|---|---|---|---|
| | X | Y | Z | X | Y | Z | X | Y | Z |
| Group MAX | 8.1 | 1.0 | 12.2 | 2.9 | 42.2 | 54.7 | 15.7 | 81.4 | 76.0 |
| Group MIN | -7.6 | -2.5 | -17.0 | -17.6 | -66.3 | 0.2 | -9.9 | -33.3 | 16.7 |
| Group ROM | 15.8 | 3.5 | 29.2 | 20.6 | 108.5 | 54.6 | 25.5 | 114.6 | 59.4 |
| GROUP STDEV MAX | 5.3 | 0.8 | 12.4 | 5.6 | 8.9 | 9.1 | 2.8 | 8.6 | 8.3 |
| GROUP STDEV MIN | 4.5 | 1.3 | 14.2 | 5.2 | 13.9 | 6.0 | 6.9 | 10.9 | 6.5 |
| GROUP STDEV ROM | 6.1 | 2.2 | 19.1 | 7.2 | 44.3 | 25.3 | 12.7 | 46.7 | 27.6 |

# APPENDIX E: KINEMATIC DATA INTERPRETATION

| DRINK | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|
| | WRIST | | | GLENOHUMERAL | | | THORACOHUMERAL | | |
| | COR | TRANS | SAG | COR | TRANS | SAG | COR | TRANS | SAG |
| GROUP AVG MAX | 7° Ulnar Dev | 2° Pronation | 7° Flexion | 4° Add | 14° Int rotation | 44° Flexion | 3 ° Adduction | 20° Int Rot | 80° Flexion |
| GROUP AVG MIN | 11° Radial Dev | 9° Supination | 56° Extension | 34° Abd | 65° Ext. rot | 9° Extension | 16° Abduction | 36 External rot | 9° Flexion |
| GROUP AVG ROM | 18° | 11° | 64° | 38° | 51° | 53° | 19° | 56° | 89° |
| **COMB** | | | | | | | | | |
| | WRIST | | | GLENOHUMERAL | | | THORACOHUMERAL | | |
| | COR | TRANS | SAG | COR | TRANS | SAG | COR | TRANS | SAG |
| GROUP AVG MAX | 20° Ulnar Dev | 3° Pronation | 12° Flexion | 3° Add | 6° Int Rot | 52° Flexion | 1° Adduction | 42° Internal rotation | 142° flexion |
| GROUP AVG MIN | 10° Radial Dev | 5° Supination | 37° Extension | 64° Abd | 94° Ext Rot | 33° Extension | 57° Abduction | 48° external rotation | 15° Flexion |
| GROUP AVG ROM | 30° | 8° | 49° | 67° | 100° | 85° | 58° | 90° | 126° |
| **REACH BACK** | | | | | | | | | |
| | WRIST | | | GLENOHUMERAL | | | THORACOHUMERAL | | |
| | COR | TRANS | SAG | COR | TRANS | SAG | COR | TRANS | SAG |
| GROUP AVG MAX | 19° Ulnar Deviation | 4 ° Pronation | 3° Flexion | 20° Add | 19° Int Rot | 4° Flexion | 4° adduction | 37° Internal rotation | 25° Flexion |
| GROUP AVG MIN | 3° Radial Deviation | 2° Supination | 42° Extension | 6° Abd | 48° Ext Rot | 46° Extension | 24° Abduction | 22° External rotation | 23° Extension |
| GROUP AVG ROM | 22° | 6° | 45° | 26° | 68° | 50° | 28° | 59° | 48° |
| **REACH ACROSS** | | | | | | | | | |
| | WRIST | | | GLENOHUMERAL | | | THORACOHUMERAL | | |
| | COR | TRANS | SAG | COR | TRANS | SAG | COR | TRANS | SAG |
| GROUP AVG MAX | 8° Ulnar Deviation | 1° Pronation | 12° Flexion | 3° Add | 42° Int Rot | 55° Flexion | 16° adduction | 81° Internal Rotation | 76° Flexion |
| GROUP AVG MIN | 8° Radial Deviation | 3° Supination | 17° Extension | 18° Abd | 66° Ext Rot | 0° | 10° Abduction | 33° External rotation | 17° Flexion |
| GROUP AVG ROM | 16° | 4° | 29° | 21° | 109° | 55° | 26° | 114 | 59° |
| | | | | | | | | | |