Medical Devices and Assistive Technology in the Treatment of Individuals with Hypermobile Type Ehlers-Danlos Syndrome (Type III)

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Abstract

Hypermobile type Ehlers-Danlos syndrome is a rare genetic disorder that it is primarily received hereditarily. Symptoms of this disorder are widespread and range from mild to severe. Their effect on the body in some individuals is hardly noticeable, however, in others it prevents the completion of the most minor of tasks. Due to the severity of symptoms that can be presented, there are multiple comorbid disorders that go hand in hand with this genetic disorder. With these comorbid disorders, there are many forms of assistive technology and medical devices that can be used to increase an individual's quality of life. With the implementation of these devices into a patient's treatment plan, the individual is granted freedoms they did not have before, as well as peace of mind in their own health treatment plan. Medical devices for individuals with hypermobile type Ehlers-Danlos syndrome vary depending on the specific need, however, they can be tools to help the gastrointestinal system, Central Venous Catheters, or surgical implants. Assistive technologies that are beneficial include mobility aids, monitoring devices, and external supports. The use of these technologies and devices are extremely beneficial to the livelihood of the patients and assist in the completion of activities of daily living that may otherwise be unattainable or unsafe for them.

Keywords: hypermobile type Ehlers-Danlos syndrome (hEDS), postural orthostatic tachycardia syndrome (POTS), subluxation, gastrointestinal, mobility, assistive technology, medical device

Executive Summary

Hypermobile-type Ehlers-Danlos syndrome (hEDS) is rare, however, can be very severe and negatively affect the quality of life for individuals who are affected. The symptoms are variable for each affected, however, they are organized into how the relate to specific parts of the body: skin, joint, bone, gynecological, neurological, gastrointestinal, and cardiac. Each type of symptom comes with it's own form of affecting the body and no two individuals will experience hEDS in the same way.

Specific symptoms an individual presents can be part of another disorder as well, a comorbid disorder, which may then have their own individual symptoms on top of the already present ones. These comorbid disorders include gastroesophageal reflux, gastroparesis, chronic fatigue syndrome, postural orthostatic tachycardia syndrome (POTS), and orthostatic hypotension. Symptoms of gastrointestinal disorders include nausea, vomiting, heartburn, and an inability to consume nutrients. Orthostatic hypotension and postural orthostatic tachycardia syndrome share many symptoms as well. In severe cases POTS can result in dehydration, fainting, dangerously high heart rate, blurred vision, and weakness. Orthostatic hypotension is signified by a drop in blood pressure which can also result in fainting. Combining all of these possible symptoms for a patient results in the need to create management plan with their doctor. There is no cut and dry way for individuals to be treated, therefore all plans are specialized and take the lifestyle of an individual into account in order to provide them with the best quality of life possible.

The management of symptoms in individuals with hEDS and the various comorbid disorders is achieved through the use of medical devices and assistive technology. Medical

devices to assist with gastrointestinal dysfunction include six different types of feeding tubes that allow an individual to get the necessary nutrition that they would otherwise be unable to achieve. Cardiac dysfunction can be treated or assisted with medical devices, such as long term intravenous lines, and assistive technology like heart rate and blood pressure monitors, as well as mobility aids. Considering the effects of joint instability on the body, widespread pain, bone density, and cardiac dysfunction; wheelchairs, rollators, and walkers can be beneficial to the individual in mitigating and prevention of symptoms. The joints and bones can also be supported with external braces, surgically implanted supports, or surgically corrected with ligament repairs. Medical devices and assistive technology can be used in conjunction or individually for an individual to treat as many symptoms as possible in order to manage the effects hypermobile-type Ehlers-Danlos syndrome has on the body.

For an individual who goes untreated, hEDS can result in them being house bound or bedbound relying on others for help to complete each and every task of living. Other individuals who are untreated are only affected in specific situations but are still not able to complete all the tasks and activities that are required of them to be a functioning member of society. Implementing in the necessary medical devices and assistive technology that are discussed in this paper allow an individual to complete the tasks that are necessary for living as well as gain back the freedom that they may have lost due to untreated symptoms.

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Introduction

Ehlers-danlos syndrome is a rare genetic disorder that affects the body systematically. While there are five different types, the third one, hypermobile type, is becoming prevalent in society. In diagnosing hypermobile type Ehlers-Danlos syndrome, also referred to as hEDS, an official diagnosis can be given only after proper genetic testing. Published in *IRSN Dermatology* (2012), this form of Ehlers-Danlos syndrome accounts for nearly 50% of diagnosed individuals and the classification comes from the lack of specific cutaneous and membrane symptoms seen in other forms. This type also represents a group of people with joint hypermobility as defined by the Brighton criteria (Castori, 2012). The Brighton criteria is a list of specific points on the body which should be able to achieve a specific position when flexed. These points are attributed to a points system. The number of points at which an individual is hypermobile are calculated in order to determine the overall level of hypermobility that person experiences. The presence of both major criterion, 1 major and 2 minor, 4 minor, or 2 minor and a affected first-degree relative are required for diagnosis. Table 1 below displays the Brighton criteria for diagnosing joint hypermobility syndrome. The Beighton score is determined by the hyperextensibility of nine different joints. These joints are as listed ("Beighton Hypermobility Score", 2017):

- 1 point for the ability to touch the floor with flat palms and straight legs, bending at the waist
- 1 point for each elbow that bends backwards
- 1 point for each knee that bends backwards
- 1 point for each thumb that touches the forearm when bent down

If an individual has 4 or more points, they meet the first major Brighton criteria. If they only get 1-3 points they are considered to have met a minor criteria.

Brighton criteria for diagnosing hEDS (Castori, 2012).

Major Criteria	Beighton score greater or equal to 4/9
	Arthralgia of more than 4 joints for more than 3 months
Minor Criteria	Beighton score of 1-3
	Arthralgia in 1-3 joints
	History of joint dislocations
	Greater than 3 soft tissue lesions
	Marfan-like habitus
	Skin striae, hyperextensibility, or scarring
	Eye signs, lid laxity
	History of varicose veins, hernia, visceral prolapse

While the cause of hEDS is known to be genetic, why it happens is still unknown and the symptoms are highly specific to each individual. As a result of this, there is no set in stone form

of treatment nor is there a cure. In understanding the symptoms of hypermobility type Ehlers-Danlos syndrome, professionals can make educated decisions regarding assistive technology and medical devices in order to provide their patients with the highest quality of life possible.

Review of Research

The nature of the research which has been completed thus far revolves around the medical diagnosis of hypermobile type Ehlers-Danlos syndrome. It is necessary to understand the range of symptoms and levels of severity which they can portray for an individual who has hEDS in order to understand how different treatments can help them. This medical portrayal also reaches out into comorbid disorders whose symptoms can be debilitating as well, but these disorders are not always present in every individual. While there are treatment and management options out there, it is not cut and dry when considering which will work best for each person. In order to develop the idea that assistive technology is beneficial in cases of hypermobile type Ehlers-Danlos syndrome, the potential symptoms and comorbid disorders must be understood in relation to how they affect an individual's quality of life.

Discussed in multiple sources, the symptoms that can be experienced because of hEDS vary widely and can be spread throughout the body with seemingly no connection. Marco Castori (2012) makes a point of the systematic nature of hEDS in section five of his article called "Clinical Manifestations". The hEDS clinical description article within the *American Journal of Medical Genetics* has all of the same categories when referencing the effects hEDS has on the body with an additional few. In using the two of these articles in conjunction, it is possible to

paint an image of all the systems which can be affected by hEDS. This image will create an understanding and a way of explaining exactly how a specific symptom can be debilitating. Boonpongmanee discussed a specific part of the symptoms that arise when an individual is diagnosed with hEDS: gastrointestinal dysfunction. This study focused on a survey completed by individuals with classical or hypermobile type Ehlers-Danlos syndrome. This survey gave an estimate of the prevalence of individuals affected by specific gastrointestinal malfunctions. While this study only focuses on one type of impairment related to hEDS, there are many more ways in which an individual can be affected.

Hakim and the group of authors with him wrote 2 articles in the *American Journal of Medical Genetics* regarding comorbid disorders. One of these was focusing on chronic fatigue syndrome (CFS) which is a disorder many live with for years before receiving a diagnosis of. The second was dysautonomia, in which they autonomic system (responsible for involuntary muscles) is dysfunctional. Specific forms of dysautonomia and their symptoms can be linked back to hEDS because a symptom of hEDS is cardiac dysfunction. It is apparent that some of the symptoms in comorbid disorders crossover with hEDS, however, they also bring in other unrelated symptoms which can cause negative effects to an individual's quality of life.

Although there are many different methods of treatment for specific symptoms, there is way to determine what will be right for each individual. There are medical devices and forms of assistive technology that can be used to help. Each person will not experience this disorder the same way, and treatment plans need to be individualized towards the specific set of symptoms that are present.

Discussion

When it comes to management of hypermobile type Ehlers-Danlos syndrome, there are several different options that can be used depending on the symptoms that are presenting themselves. Two types of treatment for hEDS that will be discussed are the available medical devices and assistive technology that can be used to increase a patient's quality of life. Medical devices are used to counteract and mitigate specific symptoms, however, they are often too invasive to be used as preventative measures. Assistive technologies are highly beneficial as a way to both mitigate symptoms as well as prevent symptoms. These two treatments can be used in conjunction or on their own depending on the needs of the individual. In order to understand why specific treatment methods are beneficial and learn which ones could be the most useful for specific situations, the symptoms of hEDS and resulting comorbid disorders must first be understood. The first step of management is a diagnosis of hEDS itself and all comorbid disorders that are present along with it, then the individual and their medical team have the ability to begin searching for the technology and devices that will provide them with the highest quality of life.

Symptoms

The symptoms resulting from hEDS are systematic in nature and effect each individual uniquely. Cutaneous features are most commonly seen with classical type Ehlers-Danlos syndrome, however, skin hyperextensibility is often seen in hEDS according to M. Castori (2012). That being said, the most defining feature of hEDS, and associated only with this specific type of Ehlers-Danlos syndrome, is called congenital capsulo-ligamentous laxity. As a result of this, the joints often have excessive movement, instability, and injuries to to the ligaments. It is

undetermined if repeated joint subluxations ultimately degrade or improve the joints as they can make the ligaments more elastic or cause them to stiffen, However subluxations come with a significant amount of pain that can cause a less of function (temporarily or permanent) for the individual (Castori, 2012). There have been reported gynecological symptoms as a result of hEDS as well. In the American Journal of Medical Genetics, it was stated that 67% of women report mucosal problems and 73.3% of women reported urinary incontinence (Tinkle et al., 2017). Neurologically, widespread pain is one of the largest complaints in individuals with this genetic disorder. Chronic or recurrent pain affects the musculoskeletal system as well as internal organs. Headaches and migraines have been noted as being common experiences, both of which can be disabling to an individual (Castori, 2012). Due to the pain that is involved with hEDS directly and as a result of comorbid disorders, there is a higher prevalence of psychological disorders such as depression, anxiety, and sleep disorders (Tinkle et al., 2017).

Comorbid Disorders

Specific symptoms of hEDS stem from the disorder itself, but can emerge as a comorbid disorder with it's own set of symptoms as well. Often times these comorbid disorders are considered to be a symptom of hEDS themselves because of how prevalent are. Gastrointestinal symptoms are highly common in individuals with hEDS. These symptoms have the ability to progress into a gastrointestinal disorder such as gastroesophageal reflux (GERD) and gastroparesis. With GERD coming in at a report level of 74% in a study of individuals under 33 years old, it is very commonly associated with hEDS, although there is no gastrointestinal criteria in the diagnosis (Castori, 2012). A study published in the Journal of Gastrointestinal Endoscopy, 23 patients with classical or hypermobile type Ehlers-Danlos syndrome were evaluated by the National Institute

of Health. In the study, the results concluded that 53% of the individual's experience GERD, while no patient experienced prior gastrointestinal perforation before the study (Boonpongmanee, et al., 2000). Symptoms associated with GERD include heartburn, regurgitation, dysphagia, odynophagia, and noncardiac chest pain (Orlando, 2000). These symptoms can be severe enough to restrict food intake and cause negative effects to an individual's quality of life. Gastroparesis -- also known as delayed gastric emptying -- is commonly experienced with nausea, vomiting, and a fullness with cyclical nature (Halser, 2012). This comorbid disorder can cause heightened pain in individuals, most notably in the abdomen area from fullness and bloating. As a result of gastroparesis, individuals affected typically consume less than 60% of daily nutrient recommendations (Halser, 2012). Gastrointestinal dysfunction is not the only part of the body affected by a comorbid disorder, chronic fatigue can negatively affect an individual's quality of life as well. Chronic Fatigue Syndrome (CFS) is categorized by inordinate levels persistent, recurring fatigue that cannot be explained by other conditions (Hakim, De Wandele, O'Callaghan, Pocinki, Rowe, 2017a). While certain conditions rule out CFS from being the cause of fatigue, it is often experienced with hEDS that it has been regarded as a comorbid disorder. Cardiovascular dysfunction is another prevalent symptom, and in severe cases, is immensely dangerous. One form of cardiac dysfunction is Postural Orthostatic Tachycardia Syndrome (POTS), a type of dysautonomia. Hakim et al. (2017b) explains the main symptoms associated with POTS are syncope, blurred vision, tachycardia, lightheadedness, and weakness upon standing. The two diagnostic requirements as discussed in the article are an increase in heart rate 30 beats per minute within 10 minutes of standing, as well as a lack of orthostatic hypotension. Orthostatic hypotension, also considered to be a comorbid cardiac

dysfunction disorder, is characterized by a drop in blood pressure greater than 20 mm/Hg when standing (Hakim, et al., 2017b). The symptoms of comorbid disorders can be just as severe as the symptoms of hDS and must be managed accordingly.

Medical Devices

There are multiple medical devices that can be used in order to help an individual mitigate their individual symptoms, however, these are often surgically implemented and are typically considered too invasive to be used unless they are absolutely needed. For individuals with severe gastrointestinal symptoms, most commonly gastroparesis, feeding tubes are necessary. The Feeding Tube Awareness Foundation discusses the six different types of tubes and how each of them is a different combination of entry position and exit position. Nasogastric, nasoduodenal, and nasojejunal tubes all enter the body through the nasal cavity and are extended into various points in the stomach and small intestine. Gastric, gastrojejunal, and jejunal tubes are placed in the abdomen and feed directly into the desired position. Gastric and nasogastric tubes exit directly into the stomach, gastrojejunal tubes have two lines extending into the stomach as well as the small intestine, nasoduodenal tubes exit at the beginning of the small intestine (just after the stomach), and lastly jejunal tubes (naso and abdominal) feed directly into the small intestine. Each type of tube is used depending on the bodily functions of the individual and the ways in which their gastrointestinal system is malfunctioning (“Tube Types”, 2017). Two things the patient has to consider when discussing with their gastrointestinal doctor about feeding tubes are how long the intended treatment time is and if there is a situation where a nasally placed tube would not be ideal. As the nasally inserted tubes have to be secured in place with tape, an individual with an adhesive allergy would not be a good candidate for this method and may be

expedited to an abdominally placed tube. For those prone to vomiting, nasal tubes do not always stay in place during the process and an abdominal tube may be a safe and time efficient route that bypasses the potential need for additional surgeries. Using a feeding tube allows for an individual to supplement their nutrient intake that has been lacking due to the inability to eat. In some cases, a feeding tube will entirely replace a patient's nutrient intake. As stated earlier by Halser (2012), gastroparesis can result in the lost of more than 40% of nutritional intake, this loss, over time, can trigger the body into starvation mode and a form of non-digestive feeding must take place. Feeding tubes are an essential form of treatment for individuals with severe gastrointestinal functioning problems.

Central venous catheters (CVCs) are long term intravenous (IV) lines that feed directly into a vein for long term medication or fluid administration (American Cancer Society, 2016). CVCs allow patients to receive treatment from home as opposed to going to infusion centers and they also allow for frequent treatments because once the line is placed, there is no manually placing an IV each time treatment is needed. Two of the most common types of CVCs that are used are PICC lines and ports. The American Cancer Society explains the differences between PICC lines and ports as well as the advantages and drawbacks to each. PICC lines are placed into a vein in the arm and end at a vein near the heart. This form of line does not require surgery for placement. These lines are able to stay in for months at a time, but there are very specific guidelines for caring for the line. A port, also known as an implantable venous access port, is permanently placed under the skin and lasts for years. When deaccessed, it does not affect the individual in any way, but it does require surgery to have it placed (American Cancer Society, 2016). Fluids are very important for individuals with POTS as they are prone to dehydration and low sodium

levels. In order to counteract the effects of this, saline fluid treatments are effective for receiving quick benefits and relief from symptoms (Koichi Mizumaki, 2011). Weekly treatments, while the most effective, can be very time consuming if they are done in an infusion center and needed to have a regular IV placed each week is stressing on the body. Placing a CVC for individuals to receive treatment allows them to receive it from home, and even on the go if necessary. Other forms of treatment given venously can vary from pain treatments to daily vitamins. For patients who struggle with very severe pain as well as gastrointestinal malfunction, IV pain medication may be necessary. There are multiple different ways in which permanently placed lines can benefit an individual, each of which must be discussed with a medical team in order to determine what the best route of care will be.

One of the most surgically intensive methods of treatment for hEDS is the use of implants or other surgical procedures to support or repair, bones, tendons, and ligaments. Wires, nails, rods, and pins can be placed into the body to treat orthopedic fractures and breaks. Individuals with hEDS are also prone to developing osteoporosis (Tinkle et al., 2017). As a result of this, they may experience more serious fractures and breaks in their bones than others. Using devices to assist in healing the bones and keeping them in the correct place is highly beneficial. Ligament reconstruction can be used for individuals who have torn or otherwise damaged a ligament. Most often, this is used in the knee or ankle, however, repairs of ligaments in the wrist are becoming increasingly popular. Scapholunate reconstruction has many techniques to achieve the desired result and was utilized successfully in 40 patients between 2009 and 2013, one of which made it back into the professional football field (Loveridge, Cutbush, Couzens, Ross,

2013). Again, medical devices and surgical measures tend to be used as a response to the presence of a specific symptom or as a specialized treatment.

Assistive Technology

Assistive technology is a broad field that encompasses a multitude of different products. Three of these product groups that will be discussed are mobility aids, external supports, and monitoring devices. External supports are easy to implement and can be cost effective for an individual to use on their own. Braces for the knees, ankles, and wrists are easy to acquire, although an individual may wish to pursue a custom fitted support for extended use. Other braces that are seen used with hEDS are finger splints and cervical collars. Finger splints prevent the fingers from subluxing while writing, using a phone, picking things up, or other activities allowing them to complete these activities with lowered levels of pain. These splints can be highly stylized and are usually made of metal. Collars are very popular for supporting the cervical spine and stabilizing the area to prevent further damage. These can be prescribed by a doctor or used to determine whether or not a spine fusion would be helpful in reducing symptoms ("Cervical Collars", 2017). Braces and supports give an individual a feeling of safety in knowing that there is an extra measure of protection keeping their joints in place. Not only that, having a brace in place greatly reduces the chance of an extremely painful subluxation from happening as is common with joint hypermobility.

Devices that are used to monitor vitals are also very helpful with this form of Ehlers-Danlos syndrome. With the occurrence of a comorbid cardiac dysfunction, heart rate monitors and blood pressure monitors are simple, yet powerful devices to use. Orthostatic hypotension is characterized by a drop in an individual's blood pressure upon standing, as mentioned earlier.

Making a patient aware of their healthy or 'normal' level allows for an individual to keep record of when their blood pressure is dropping and find ways in order to work around that. One result of dangerously low blood pressure is syncope, or fainting (Hakim et al., 2017b). If using a wrist monitor, it is possible for an individual to detect when they are going to have a syncopal episode and attempt to avoid it as much as they can. POTS is similar to orthostatic hypotension in that one of the most serious, yet common, symptoms is tachycardia induced syncope (Castori, 2012). Using a wrist based heart rate monitor allows constant recording of one's beats per minute and real-time reporting for an individual to base their actions off of. Other popular monitors include using finger based and phone based devices to periodically track their heart rate. These do not allow for constant recording, but do allow for quick check in's if an individual is beginning to feel symptomatic. Using a heart rate monitor allows an individual to judge exactly what physical efforts will be too intensive for them depending on their current heart rate or if they need to cease their activity for safety reasons. Other types of medical devices may help alleviate the need for strict monitoring of blood pressure and heart rate, as well as assist other dangerous symptoms. Physical movement and functioning can be extremely taxing on hypermobile individuals. With the widespread pain that is experienced, fatigue, subluxations, dysautonomia, and more, mobility devices are becoming increasingly popular. A very common aid that is used for mobility are wheelchairs. By allowing an individual to stay seated, the fall risk associated with syncope due to POTS or orthostatic hypotension lowers, and less weight is put onto the lower body, reducing the risk of subluxing below the hips. Using wheels as opposed to walking reduces exhaustion and pain associated with the action. A device called the SmartDrive MX2 + PushTracker is a small motor that can turn manual wheelchairs into powered ones, controlled by a remote control

("SmartDrive® MX2+PushTracker™", 2017). Some patients with hEDS are affected by upper body subluxations as well as lower, meaning, manually pushing a wheelchair can be just as taxing and painful as if no wheelchair was used at all. Utilizing the SmartDrive device allows these individuals to power their chair with less of their own strength, causing less pain and more independence. Two other forms of very heavy weight bearing mobility aids are rollators and standard walkers. Both of these allow the individual to place their weight onto the device instead of their back and feet reducing pain, fatigue, and various other symptoms. While walkers need to be picked up or slid along the ground, rollators have 4 wheels allowing them to be smoothly moved over various terrains. Using these forms of assistive technologies can be highly beneficial in allowing an individual to become more independent.

For individuals who do not require the level of assistance a wheelchair, rollator, or walker provides, forearm crutches are an alternative. A cane can be very useful for providing balance and minimal weight bearing assistance, however, the wrist becomes very stressed and for an individual who may have hypermobile wrists, this is a cause of aggravation and potential pain. Forearm crutches on the other hand, allow for the weight to be distributed to the forearm or elbow to reduce the amount of pressure that is placed on each individual joint. The StrongArm Forearm Cane is a lightweight simplistic design for users that features a standard cane with an extension that wraps around the forearm and supports it from underneath (“Products”, 2017). The design of this allows for either 1 or 2 to be used at a time depending on the individual need. Another brand of forearm crutches is the smartCRUTCH. This design allows for the armband and handle portion to be rotated to find the most comfortable fit, and it allows for the angle of the forearm part to be adjusted so that the location of the pressure on it can be changed (Roux,

2016). Forearm crutches are often times more socially acceptable than a cane for the younger population with hEDS, especially with the different colors and patterns that are now available. Another beneficial mobility aid for individuals is devices that help to reach objects. With chronic pain and fatigue comes the decision of using energy for something small or saving it for something larger later. A reaching aid allows for an individual to pick up things they have dropped, or objects that are too far away, without having to go through the physical stress of moving or bending. For individuals who use wheelchairs, the amount of energy it takes to transfer from their current location and into their chair just to move 4 feet over to get the TV remote can be extremely taxing. A reaching aid would eliminate the need to transfer and use wheels by simply allowing them to grab items from their current location. Giving the individual a way to achieve small tasks without the use of extra energy allows them to take the opportunity to use that energy later for an important task or activity.

Conclusion

Hypermobile type Ehlers-Danlos syndrome is a rare, yet debilitating genetic disorder. The symptoms it produces are highly varied from individual to individual and systematically affect the body. The variability of this disorder presents itself to a lack of treatment guidelines and struggles in discovering what treatments are correct for an individual based on the symptoms they experience. There are a multitude of medical devices and assistive devices that allow individuals to determine the best course of treatment and decipher what methods work the best for them. Surgically implanted medical devices including feeding tubes central venous catheters, bone or joint supports, and ligament reconstruction are all viable options for severe situations that cannot be otherwise treated successfully. Assistive technology allows the patient to utilize

mobility devices, health monitors, and reaching devices to lessen their energy output, lowering the risk for symptoms and gaining independence for themselves. When it comes to managing the symptoms of both hEDS and the present comorbid disorders, it is clear how disabling hypermobile-type Ehlers-Danlos syndrome can be and how it can result in a lower quality of life. With the use of implanted devices as well as external aids, an individual can get the most out of their body and learn how to work with their disorder, not against it.

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Appendix