March 26, 2013

Client:

D.O.B:

Diagnosis: Ehlers-Danlos Syndrome hypermobile type

Equipment Requested: Permobil M300 Corpus 3G

To Whom It May Concern:

I am writing regarding Allison, a 13-year-old young woman diagnosed in January 2013 with Ehlers-Danlos Syndrome (EDS) hypermobile type through genetic testing. Ehlers-Danlos Syndrome is a genetic condition that affects 1 in 20,000 individuals. Ehlers-Danlos hypermobile type affects 1 in 5,000 individuals. Individuals with EDS have a defect in their connective tissue, which is the tissue that provides support to many body parts including the skin, muscles and ligaments. This defect in connective tissue presents as faulty collagen. Collagen is a protein that acts as “glue” in the body to add strength and elasticity to connective tissue. This faulty collagen results in fragile skin and unstable joints throughout the body.

Common features of EDS hypermobile type include joint hypermobility and frequent dislocations. Due to joint hypermobility, individuals with hypermobile EDS also have an increased chance to have sprains or dislocations as well as flat feet. They may also experience fatigue, muscle cramps, or early onset arthritis related to the underlying looseness of the joints. Other findings may include smooth velvety skin, easy bruising, and an increased chance for hernias. Musculoskeletal pain has the largest impact on the quality of life for individuals with EDS and this can increase in frequency and severity as individuals with EDS age. Many may develop constant and chronic pain. Fatigue can also be an associated complication. Additionally, sleep problems are commonly reported for individuals with EDS, likely due to chronic pain and depression or anxiety.

Allison is currently enrolled in the 8th grade at \_\_\_\_. Allison was previously in the academically gifted program, but due to her recent medical complications, joint pain and chronic dislocations, Allison has missed an increasing amount of school and has been pulled out of this program. Allison remains in school and accommodations are being made to facilitate the best possible learning environment for her. Allison is still very much a bright, social 13-year-old girl with many friends. When able to, Allison enjoys hanging out with her friends both in and outside of her home. Allison lives at home with her mother and her older brother, who has Ehlers-Danlos syndrome type IV, vascular type. He requires a great deal of medical attention also. Allison and her family live next door to their extremely supportive grandparents, who recently gave them the condo that is adjacent to the one that they live in.

Allison and her family have recently been approved for ‘Welcome Home Angels’, which will be providing much needed modifications to their home including a ramp for safe wheelchair entry in to the house, a wheelchair accessible bathroom, a bedroom designed specifically for Allison and an accessible kitchen, along with increased living space for the family. Allison has also been approved by CAP. The public bus system, which includes handicapped accessible busses, has routes to and from their home that will allow Allison to get to school and doctor appointments while modifications are made to their family vehicles. Due to the acute onset of Allison’s situation, options for these vehicle modifications are being explored concurrently with her home modifications and power chair attainment. The family has a truck that they will be able to adapt with a ramp to transport her chair, but also are exploring the option of having a lift put on their other SUV. Eventually when they are able to obtain the appropriate funds the family hopes to acquire an adapted van to help them transport the power wheelchair.

Overall, this diagnosis is very acute. But, it has been rapidly progressing and has taken her from walking to power mobility over just a few months. Allison’s medical and functional futures are uncertain at this time but every effort is being made to provide support for Allison to make her as functionally independent as possible.

Impairments and limitations that affect Allison’s functional independence:

*Strength*: Generalized weakness is found throughout her body. Allison does not have antigravity strength in her upper or lower extremities. She is noted to pick up her legs with her arms when she is trying to clear a footrest to transfer. Due to her global limits in available joint ranges of motion, her muscles are put at a mechanical disadvantage to generate forces. Forces that she does generate appear to cause her discomfort as they create imbalances at the joint when they contract because her joints lack connective tissues and are unable to block any intraarticular movement that her muscle contractions create. Allison has functional strength in her arms but is very limited in what she is able to do by her potential to dislocate her joints, especially her shoulders. Allison is currently learning how to perform isometric contraction exercises in an attempt to preserve the muscle strength that she currently has in her available ranges.

*Balance*: Allison is able to sit unsupported in a chair. She is unable to maintain sitting balance with anything greater than moderate balance perturbations. She does not lean while in her chair or reach outside her base of support due to joint pain and fear of joints moving. When standing, Allison’s functional balance greatly decreases. Allison is unable to maintain standing without support from one or two of her arms. With an upper extremity support in standing, global shaking and trembling are noted as Allison works to maintain her balance. Allison reports that she feels unsteady when standing, like she is ‘hurts and she is falling a lot’. When she stands she weight bears about 75% of her weight through her right leg, causing increased right hip pain. Allison has fallen numerous times when she has attempted to walk more than a few steps, putting her at high risk for further injury. Allison is able to perform an upper extremity task (brushing her teeth, washing face) while standing at a chest high table with both elbows supporting her, freeing her hands.

*Transfers*: Allison comes to stand from sitting through extending her lower back and pushing her pelvis up before the rest of her lower body. Allison reports that she has to do this because if she does not, her spine feels like it is grinding, unstable and painful. This method is particularly unsafe for her because as she contracts her muscles she is putting undue stress on her low back, pelvis and femur, causing shifting in her joints due to instability and unopposed muscle forces. Allison is noted to lurch forward with her upper trunk in an attempt to get her center of mass over her base of support in standing. When she does go to stand there is audible popping. Allison has a history of dislocating various joints during transfers. Allison is unable to be assisted in transfers from someone because of the undue pressure it can put her body wherever that person were to place their hands. Allison has ribcage instability so she is unable to tolerate anyone trying to help her by supporting her thoracic region. Allison also lacks the shoulder range of motion to put her arms around someone’s neck to help with a transfer.

*Gait*: Allison is unable to walk more than a few consecutive steps due to her pain and instability. Her observed gait has been unstable and unsteady. Allison uses various supports in her environment to support herself with her arms while she moves. Her steps are short and her base of support is widened to provide herself with more stability. Audible popping and clicking of bilateral hip joints can be noted during her gait. Allison is unable to use a walker while walking to improve her stability due to the increased stress it causes at her wrist and shoulders. Allison has the grip strength required to hold a walker, but cannot tolerate the end range wrist extension that it demands due to potential for dislocation and pain. Also, when attempting to use a walker, Allison’s shoulder instability allows her humeral head to be pushed against her acromioclavicular joint when any weight is put through her upper extremities, causing pain from bony contact and also jeopardizes the well being of her rotator cuff tendon by increasing her potential for impingement.

*Wheelchair mobility*: Allison is unable to independently use a manual wheelchair due to her risk of shoulder dislocation when self-propelling the wheelchair because self-propelling a wheelchair requires Allison to move her shoulders past 30 degrees of shoulder extension. Allison’s shoulders dislocate when moved past 30 degrees of movement any direction (shoulder flexion, abduction, extension, horizontal adduction, internal rotation and external rotation). Allison has dislocated her shoulders numerous times in the past few months when attempting to self propel the manual chair that she had been using in her transitional period from diagnosis until recently when she was able to obtain a loaner power wheelchair, discussed below.

*ADLs*: Currently, Allison is somewhat independent in her activities of daily living, or ADLs, but in a manner that puts her at great risk for falls and other injuries or complications. Allison requires help to get in and out of the shower but is able to wash herself while standing in the shower by supporting herself with her arms throughout the process. Allison is put at grave risk for slipping or falling in the shower with the wet, slippery surface combine with her instability in standing. Appropriate shower seat options are currently being explored for Allison. Allison is able to move about her kitchen at home to prepare food by scaling along the counter. She will lightly lean on the counter while she prepares herself food with her upper extremities. Allison is able to feed herself independently but is unable to prepare food or get food out for herself because she cannot tolerate the weight of most items that she needs (milk jug, etc). She is also unable to open the packaging of foods. Allison dresses herself lying supine in bed. This process takes her a long time and can sometimes cause increased pain as she progresses. Allison struggles opening the drawers in her room to get her clothes out due to the weight of the drawers and the stress it puts on her wrists and hands. Allison currently is independent with her toileting needs.

Recently, Allison obtained a loaner Permobil C300 powerchair. This powerchair has provided her with independence in mobility, as Allison has been able to navigate her school and classroom environments without assistance for propulsion or steering. This powerchair chair has been especially helpful for Allison since her recurrences with shoulder dislocation in a manual chair. She has displayed the ability to use the various functions on the chair to make her environment more accessible, including:

* Using the seat elevator to arrange herself at a classroom table so that her upper extremity joints (shoulder, elbow, hand and fingers) are in the best position to use her tablet, paper and/or pencil.
* Using the tilt function to provide pressure relief during the day and to obtain pain relief with a change of position for her hip joints.
* Using the leg rest elevator to properly position her body in a way that is most comfortable and puts the least stress on her joints.
* Using the steering features to navigate classroom environments to get to her adapted tables and safely entering and exiting the elevator at school to allow her to reach classrooms.

Equipment justification:

The Permobil M300 Corpus 3G is the appropriate device for Allison, as it will provide her with much needed functional independence and safety. She has adequate mental capacity and upper extremity function to operate the chair safely and effectively. This chair will allow her to navigate and interact with her environments, independently take care of herself and will also help to preserve the functional abilities that she currently has including the ability to transfer to standing.

The M300 is a compact mid-wheel drive power wheelchair base with programmable electronics, adjustable wheel suspension, a patent-pending climbing-traction link system, and quiet motors. Its large casters provide a smooth ride and better navigation over obstacles and thresholds compared to the typical mid-wheel drive base. The tight turning radius provides excellent maneuverability. The Corpus 3G is a modular, ergonomic seating system built to accommodate changes and/or growth. The seat depth grows from 14"-22", and the width grows from 17-23", without the need to change the seat frame. The backrest is also height adjustable (20”, 23”-28”). This allows a custom fit for the specific individual, and provides accommodation for growth, orthopedic deformities, and significant weight gain or loss. The Corpus 3G can accommodate a variety of power seat functions, including Dura Core tilt, shear reducing recline, power articulating elevating legrests, and vertical seat elevation. These functions can also easily be added in the future as medical needs change. This chair was chosen after consulting with an experienced equipment expert who prefers to work with Permobil as they find them to be a reliable company with quality products. The adjustability of this seating system, as well as the support that it will offer Allison, is why it is the appropriate choice. Allison has been fit for the Permobil M300 Corpus 3G and has been able to successfully demo and utilize all of its functions.

Equipment Specifications:

The following components are also essential for Allison to increase her independence and decrease her functional limitations:

1. **Power adjustable seat height** - The power adjustable seat height allows vertical adjustment of the seat height by the wheelchair user. Elevation increases reach and provides independence with ADLs. It promotes safety with and improved independence with lateral transfers by allowing a level transfer or transfer from a higher to lower surface, which is gravity-assisted. This will allow Allison to make herself the same height or higher than the surface to which she is transferring to (car, bed, tub transfer bench). Level transfers are much easier for Allison to complete independently as they allow her to transfer to a surface without standing and having to take steps towards a target, which can be unstable and painful. Allison performs these lateral transfers throughout her day both at home and at school to beds and raised mat tables to alleviate her pain through laying and resting. This function also facilitates forward transfers by allowing legs and hips to be more extended, thereby lessening the strength required for the Allison to perform a stand-pivot transfer. This is especially important for Allison because it provides her with much needed hip extension so that she does not have to generate muscle forces so large that they disrupt hip joint, SI and low back joint integrity when going to stand. Power seat elevation also allows the user to have eye contact with others and reduces cervical strain and pain. Vertical rise also provides psycho-social benefits of being on peer level and speaking eye-to-eye. This feature will also allow Allison to adjust her height for a variety of functions including to see herself in the mirror to perform ADLs and to adjust herself at a table in order to feed herself.
2. **Expandable Controller R-net and Harness** - this is the power module located in the base of the chair that allows the input device to communicate with the drive motors and gearbox. The expandable controller is used in conjunction with an upgraded joystick. An expandable controller is also required when any alternate drive controls are being used on the power wheelchair. With this device the expandable controller can accommodate up to six different types of drive inputs. The harness secures the controller to the wheelchair.
3. **Joystick Mount Right: Swing Away, R-net & VR2** – The swing away joystick mount allows the joystick to swing in or out at any angle to allow closer access to tables, desks, and counters. It can also facilitate forward transfers by safely moving out to the side. The joystick can also be placed at any angle for appropriate hand access. The R-net Remote Color Screen Joystick is a proportional upgraded joystick that has programmable electronics with separate drives and switch options available to safely meet different access, environmental and terrain needs. Mono jack ports will allow specialty switches and controls to be used to operate the on/off and modes/profiles function. This is needed when the standard push or toggle buttons are not accessible due to lack of activation strength or limited active range of motion.
4. **Multiple Seat Function Control Kit for R-net** - The Multiple Seat Function Control Kit describes the electronic components that allow the user to control two or more of the following actuators from a single interface: power wheelchair drive, power tilt, power recline, power leg elevation, and power seat elevation. It includes a function selection switch, which allows the user to select the motor that is being controlled and an indicator feature to visually show which function has been selected. Allison will be able to use the tilt function for pain relief, foot clearance and to keep her back in her seat when she is not able to do so independently. Allison has found the tilt function to greatly help improve her comfort level in her current trial power chair. This is incredibly imperative for Allison because she has struggled with finding positions to decrease the amount of pain she is experiencing at a certain time. Frequently Allison has found that hip extension is the only movement that can bring her pain relief. It can also be used for pressure relief, which will be imperative for Allison since she will be sitting in her chair throughout the majority of her day as she currently does, and this will put her at a higher risk of pressure ulcers or other complications. Allison experiences substantial hip pain when sitting upright and the recline function will help give her comfort by allowing her to change the angle between her back and pelvis throughout the day. All of these functions will allow Allison a means for gravity-assisted positioning. This is important for her to independently be able to achieve and maintain proper positioning. For example, she is not always able to scoot her hips all of the way to the back of the chair without the use of tilt. These functions will also provide enhancement of function. For example, using the tilt function to tilt herself anteriorly may be clinically beneficial to assist with functional reaching. Reaching is a struggle for Allison due to her limited range of motion, so any additional help she can get will allow her to remain more functionally independent. Allison will also be able to use the tilt function to allow her to carry objects in her lap since she is not able to do so with her upper extremities due to her strength and range of motion deficits. Anterior tilt can be used with seat elevation to improve transfers to and from elevated surfaces. This will be important for Allison because she will not always have control over the height of where she needs to transfer. This will allow her to make adjustments and be able to interact with a wider range of environmental barriers. Use of the power leg elevation and footrests can help Allison to improve her balance during completion of ADLs by allowing her to maintain a safe position during braking.
5. **Corpus 3G Ergo Back** - this is an ergonomic contoured backrest and is a component of the Corpus seating system. Aside from providing comfort, it accommodates unstable trunk positions secondary to decreased trunk and upper extremity coordination. This will decrease the amount of stress placed on Allison’s postural musculature and the forces they generate on her vertebral column. Standard back upholstery and linear seating orthosis are inadequate for postural support due to Allison’s instability, discomfort and chronic pain, and scoliosis. The recommended orthosis simulates the contours of the trunk and provide stability for positioning and can reduce the risk of Allison developing further spinal deformities. This is especially important as her condition makes her ineligible for any surgical procedures, making it a priority to prevent any further spinal deformities. The backrest is customizable upon ordering, and is further adjustable with the use of postural supports and in conjunction with seat functions. The contoured backrest will also be useful in conjunction with the tilt function of the wheelchair, as it will allow her to maintain the appropriate contact with the chair as it moves.
6. **Headrest Corpus 3G Back** **with adjustable and removable hardware**– This headrest will protect Allison’s cervical integrity as she uses the tilt and recline functions of the wheelchair.
7. **Power Elevating Center Mount Legrests** - Power articulating elevating legrests allow legrest elevation and articulation, which provides leg extension while elevating. These legrests can improve circulation and reduce or prevent edema (when combined with tilt/recline. This function will be important for Allison due to her decreased mobility and lower extremity stasis. These legrests can be adjusted to provide comfort and will allow Allison to move her lower extremities through her available range without unnecessary muscle contractions causing joint instabilities, especially hamstring contractions. Additionally, these legrests can improve ground clearance to navigate thresholds and slopes.
8. **Power Transfer Legrest – Corpus 3G** – Power transfer legrests bring the users feet down to ground level and then also raise up to help with positioning and clearance. This function is imperative for helping Allison go from sitting to standing without undue stresses placed on her body. Without this function, Allison has to pick her legs up off of the legrests and transfer them to the floor. When Allison tested this power function in her demo chair she was noted to transfer to standing more efficiently and with less instability and risk. It is imperative to preserve what functional ability that Allison currently has to transfer from sitting to standing, as it provides her with independence along with the benefits of standing including increased mobility around small areas and the health benefits of weight bearing.
9. **Medical necessities bag and hooks** – this hardware is used to secure a medical necessity bag to carry essential items such as emergency medications, catheterization/toileting supplies, etc. The clips lock to provide a safe and secure means to transport medical items. Considering that Allison’s medical status has been progressing, these may all be needs of hers in the future. Currently, Allison will require this bag to hold snacks, food and water for herself. Allison is not able to eat most food that is offered at school due to its weight, making it so that she cannot pick it up without causing pain. Allison brings appropriately selected food so that she is able to get the nutrients she requires throughout her day. She also has to carry small water bottles with her because she is not able to drink out of larger ones because of their weight. Allison also brings personal belongings with her and would use this bag to put them in.
10. **Armrest pouches** – this is a medical necessity pouch that attaches to the armrest allowing the user to carry necessary items that must remain within reach. These will be especially imperative for Allison as they will provide her the opportunity to keep something in her limited reach. Otherwise, she would have to carry all belongings on the back of her chair and she is unable to reach that independently. Of importance, Allison will need to have a cellphone near her at all times incase a medical emergency would arise. She needs access to one of these in the instance that she needs help. She is unable to reach around to anything behind her chair, and has dislocated both shoulders when trying to do so in the past.
11. **Upper Extremity Support Angle** – to support upper extremities in a neutral position to prevent shoulder subluxations and promote increased function.
12. **Bodypoint Monoflex Medium Chest Support Belt Kit** – this is required for anterior trunk support and postural stability due to lack of trunk control. This will be necessary for Allison especially when her chair is not in tilt. Allison has very delayed righting reactions of her trunk and would be at risk of falling if she were to lose her balance.
13. **Push Handles for Corpus 3G Seat** – this feature will allow a caregiver to manually push the wheelchair in the event of a chair malfunction or battery failure.
14. **Batteries – Group 34, 12V 60 Amp Hours** – this is required for the wheelchair to function without difficulty for an extended time.

15. **Sunrise Medical Jay Ion Cushion** – this cushion will provide Allison with the pressure relief, positioning and comfort needs that she has. This will be crucial to prevent any complications that can potentially arise from her prolonged sitting. The cushion also has a breathable material to help with temperature control, which will be very important for Allison as she will be spending a lot of time in her seat in a multitude of environments and climates.

Allison has had the opportunity to trial a very similar Permobil wheelchair that had many of these functions for one week. This demo was done only at school due to her house not currently being accessible for a power wheelchair. Her house will soon be under renovation to make it more accessible. A company that is very familiar with the building standards for wheelchair accessibility and safety is doing the house renovations. Allison found this Permobil demo chair to be much more comfortable than her current loaner chair and also found the functions to be very beneficial. The chair had all functions except for the power transfer legrests, so Allison was able to have increased functional independence but did still struggle with her sit to stand transfers.

The Permobil M300 Corpus 3G mobility device meets a variety of mobility and positioning needs for Allison while greatly increasing her functional independence. It will sufficiently give her the freedom and comfort that is so imperative to her, prevent deformities and provide her with much needed safety. Subsequently, this will decrease the amount of medical intervention that Allison could potentially require if she does not receive an appropriate seating device.

A team comprised of the physical therapist, wheelchair vendor, patient and her family were involved in the decision making process regarding this equipment. If you have any questions, please do not hesitate to contact us at 910-616-4424. Thank you for your assistance and consideration in this very important matter.

Sincerely,

Amy Woodcock, PT