Immediate effects of foot orthoses on gait and balance in boys with Duchenne Muscular Dystrophy: a pilot study

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Study Design: Single group, repeated measures

Purpose: To examine the immediate effects of the use of custom-made foot orthoses (FO) on gait and balance in boys with Duchenne Muscular Dystrophy (DMD) who are still able to walk independently.

Background

Duchenne muscular dystrophy (DMD) is an x-linked recessive disorder that leads to progressive muscle weakness and loss of independent ambulation by a mean age of approximately 10 years.[1,2] Loss of ambulation results from a combination of proximal leg muscle weakness and ankle plantar flexion contractures.[3] Boys who are in the middle and late ambulatory stages of DMD typically assume a posture of ankle equinus, knee hyperextension, hip flexion, and increased lumbar lordosis to maintain stability in standing and during the single-limb support phase of gait.[4,5] This posture reflects compensation for lower extremity weakness and ankle plantar flexion contractures, and promotes bilateral hip and knee extension by maintaining the line of gravity posterior to the hips and anterior to the knees.[4,5]

Quadriceps insufficiency is a key factor in gait deterioration in DMD.[5] As the knee extensors become progressively weaker, boys with DMD rely increasingly on active ankle equinus bilaterally to keep the line of gravity anterior to the knees, thereby establishing passive knee stability.[4] Ankle plantar flexion contractures continue to progress, decreasing the base of support and making balance precarious.[4] The ability to ambulate is lost when lower extremity weakness and plantar flexion contractures become so severe that effective compensation is no longer possible.[3,4]

Prolonging independent ambulation in boys with DMD can slow the progression of scoliosis[6-8] and avoid or postpone many of the other problems associated with wheelchair confinement in this population, including obesity, disuse atrophy, osteoporosis, pathologic fracture, pressure injuries to skin, and severe hip and knee flexion contractures.[4] Psychological benefits, such as increased self-sufficiency and self-confidence, also have been reported if independent ambulation can be maintained.[4,7]

Use of bilateral knee-ankle-foot orthoses (KAFOs) is sometimes instituted as a means of prolonging independent ambulation in boys with DMD.[3,6,9,10] The locking knee joints of the KAFOs substitute for weakening knee extensor musculature.[4] In a review of seven studies of orthotic intervention using KAFOs for this population, Bakker et al[6] reported that the median values for the duration of independent walking, assisted walking, and standing after intervention were 24 months, 36.2 months, and 50.5 months, respectively. However, orthotic intervention with KAFOs may not result in walking ability that is truly functional in terms of speed and distance walked.[3,6] In addition, this intervention is associated with significant drawbacks. Lower extremity orthopedic surgery, particularly Achilles tendon lengthening, is often needed for successful fitting of KAFOs.[6] The pain and expense of surgery and the risk of surgical complications are major considerations for boys with DMD and their families. Furthermore, the KAFOs themselves may be uncomfortable, difficult to don/doff, and aesthetically unacceptable.[3,6]

Use of nighttime ankle-foot orthoses (AFOs), in conjunction with passive stretching regimens, is recommended to slow development of ankle plantarflexion contractures[4, 11-13] and to enhance walking ability[14] in boys with DMD. Daytime use of AFOs, however, should be avoided because the AFOs compromise ambulation by blocking compensatory ankle plantar flexion during single-limb support and placing excessive demands on the knee extensors during the loading response.[4] Townsend et al[11] reported that declines in both gait speed (measured using the 10-m walk test) and walking capacity (measured using the 6-minute walk) were seen when boys with DMD ambulated while wearing dynamic response AFOs. In addition, parents reported an increased incidence of falls in 2 of 3 study participants.

Another possible orthotic intervention for boys with DMD is the use of custom foot orthoses with heel lifts. Based on our literature review, the use of foot orthoses with heel lifts has not been reported as an intervention for boys with DMD, and is not part of the standard of care for this population. Foot orthoses do not restrict ankle movement. They fit entirely within the shoes, and so are not visible in the types of (supportive) footwear typically worn by ambulatory boys with DMD. A review of the literature revealed a scarcity of studies on the effects of these types of foot orthoses on gait and balance in individuals with neuromuscular disorders, and no studies involving the DMD population. Foot orthoses with heel lifts should assist in supporting the ankles in plantar flexion, thereby helping to maintain the line of gravity anterior to the knees. These orthoses also should increase the weight-bearing area on the plantar surfaces of the feet, thereby increasing the base of support and improving balance.

One recent pilot study examined the effects of custom lower limb orthoses on balance, walking, and quality of life in individuals with fascioscapulohumeral dystrophy.[15] In this study, participants were tested once, at baseline, wearing their customary footwear and again, at one month after receiving their bilateral custom orthoses, wearing their customary footwear plus the orthoses. Ten of the 15 participants in the study were fitted with foot orthoses, and five received AFOs. The researchers found that both foot orthoses and AFOs were associated with improved walking, balance, and quality of life in this sample. These results should be interpreted with caution, as this was a pilot study, with no control group, no consideration for possible practice effects, and no blinding of testers. In addition, the results may not be applicable to boys with DMD, as fascioscapulohumeral dystrophy has asymmetric and highly variable patterns of muscle weakness that are quite distinct from those observed in boys with DMD.

A final motivation for exploring foot orthotic intervention for boys with DMD comes from the clinical experience of two members of our research team. In clinic, we noticed dramatic effects of foot orthoses that had appropriate heel lift for a patient who demonstrated the same impairments seen in patients who have DMD. This patient had an equinus deformity at both ankles. Her knees moved rapidly and in an uncontrolled fashion into hyperextension during the single-support phase of gait, and her gait speed was slowed by the whole profile. Foot orthotic intervention produced dramatic improvements in her gait pattern. The foot orthoses were very enthusiastically adopted by the patient, who reported being able to go hiking with her family and perform other activities that were extremely difficult or impossible without the orthoses.

Procedures

This pilot study is currently underway as of April 2017. The following discussions will report data on the first two subjects only.

1. Recruitment

Potential participants for this study are recruited by members of the research team either in person at MD and pediatric clinics, or by responses to flyers placed in these clinics (**Appendix 1**). Healthcare providers in these clinics have been provided with a list of eligibility criteria in order to help identify appropriate potential participants. While attending MD clinics in person, research team members hand out flyers about the study to healthcare providers and prospective participants, encouraging the patients/family members to call and sign up for the study. Interested parties are provided contact information for Vicki Mercer, PT, PhD.

2. Screening

Participant screening is done by members of the research team either in person or over the phone. Research team members conduct screening that is guided by a script of questions (**Appendix 2**). Eligibility criteria include: diagnosis of DMD; English-speaking/reading, able to ambulate at least 100' without assistance from another person, and walks with forefoot strike. Children who are deemed appropriate for the study will schedule an appointment at University Physical and Occupational Therapy in Hillsborough, NC for informed consent, orthotic fabrication, and completion of data collection.

3. Study Protocol

Overview:

- Custom Orthosis fabrication
- MMT Measurements
- PROM Measurements
- Balance: North Star Ambulatory & GAITRite® Assessment
- Data Analysis

Custom Orthosis Fabrication

Dr. Michael Gross, PT, PhD, is an expert in lower quarter screening and custom orthotic fabrication, and conducts the process of fabricating and fitting orthoses for the participants. Dr. Gross begins the process by examining the participant for structural malalignment in the lower extremities, noting such alignments as midfoot arch structure, forefoot to rearfoot alignment, available passive ankle dorsiflexion, and limb length inequality. He molds foot orthotic material to the participant's feet while they are in the seated position. Semi-rigid orthotics are then fabricated using thermal cork and 0.375 mm thick nickleplast (Alimed Corp, Dedham, MA) for posting and heel lift.[16] The key component to these orthotics are the heel lift using the 0.375 mm thick nickleplast, which are used to address the participant's limited passive dorsiflexion. Dr. Gross determines the appropriate amount of heel lift for each child depending on the degree of ankle dorsiflexion restriction. After fabrication, the child will test the orthoses by wearing them during ambulation, and Dr. Gross modifies the orthoses for optimal comfort of the participant.

MMT Measurements

Participants are seated in short sitting with hips and knees flexed to 90°, then asked to extend one knee through the full range of motion. If the participant is unable to perform the movement through the full range, then the knee extension lag is recorded. If he is able to achieve full knee extension, then strength testing is done using a handheld dynamometer (*Chatillon CSD 400 Dynamometer*). Isometric knee extensor muscle torque is measured with the knee at 90° of flexion and distance from the knee joint center to just proximal to the malleoli is measured in order to determine torque. Three trials are completed and the average is recorded. Strength norms from the literature[17] are used to compare participants to age-matched, typically developing peers.

PROM Measurements

Measurements using standard goniometry are performed on each participant bilaterally for the following passive ranges of motion: hip extension, knee extension, and ankle dorsiflexion. The tester instructs the participant to relax and not assist with movement of the test limb. The better of 2 trials is recorded for each lower extremity.

Hip extension is measured with the participant lying in the prone position. The tester begins by identifying the greater trochanter of the femur as the reference point for the axis of rotation. The stationary arm of the goniometer is aligned parallel to the midline of the pelvis and the moveable arm is aligned parallel to the midline of the femur using the lateral epicondyle for reference. The tester passively extends the participant's hip until maximal extension is reached, determined by a firm end-feel, and the tester records this degree of motion.

Knee extension is measured with the participant lying in the supine position. The tester positions the hip and knee in 90° of flexion. The lateral epicondyle of the femur is identified as the reference for the axis of rotation. The stationary arm of the goniometer is aligned perpendicular to the floor or parallel to the midline of the shaft of the femur. The moveable arm is aligned with the lateral midline of the fibula using the lateral malleolus and fibular head for reference. The participant's knee is passively extended until a firm end-feel is met and the tester records this degree of motion.

Ankle dorsiflexion is measured using two methods: one with the participant in prone with the knees extended and feet hanging off the test table and one with the participant in short sitting with the knees flexed to 90°. In both positions, the axis of rotation is determined by identifying the lateral aspect of the lateral malleolus. The stationary arm of the goniometer is aligned with the lateral midline of the fibula using the lateral malleolus and fibular head for reference. The moveable arm is aligned parallel to the lateral midline of the 5th metatarsal using the base and head of the 5th metatarsal for reference. The tester passively dorsiflexes the participant's ankle to maximal dorsiflexion and a firm end-feel is felt and record this degree of motion. Discrepancies are attributed to the proximal heads of the gastrocnemius placed in a stretched position when the participant is lying prone and are indicative of soft tissue tightness or contracture.

Balance: North Star Ambulatory Assessment & GAITRite® Assessment

After the foot orthoses have been fabricated, the participant completes tests of standing balance (standing with feet together, right single limb stance, left single limb stance), three trials each, with each test timed to a maximum of 20 seconds. In addition, the North Star Ambulatory Assessment is a functional measure for ambulatory boys with DMD (**Appendix 3**) that is used to measure functional performance. These tests/measures assist in characterizing the study sample and obtain information concerning the characteristics of boys who do or do not benefit from orthotic intervention. Standing balance and walking tests are completed under two conditions, with and without foot orthoses. The order of testing is counterbalanced across participants, such that half of the participants first perform the tests while wearing their customary footwear without the orthoses, and the other half first perform the tests while wearing foot orthoses.

The GAITRite® instrumented walkway (*CIR Systems, MAP/CIR Inc., Havertown, PA 19083*) is used to obtain spatiotemporal gait measures such as step length, base of support, and cadence. Participants complete two passes on the walkway in order to obtain this information. Each participant completes a total of four passes: 2 while wearing orthoses, and 2 without the orthoses.

4. Data Analysis

Descriptive statistics are calculated for balance and gait measures. GAITRite® Systems software provides values for various parameters during gait under the two conditions.

Results

As this pilot study is currently underway, data is reported for the first two subjects only.

Participant A was a 9 year-old male standing 4' 4'' tall and weighing 73 pounds. He was diagnosed with DMD in 2011. His family initially reported that he wears a size 1 shoe, but he presented to the clinic wearing a size 2 shoe. Upon measurement with a standard shoe-sizing chart, it was determined that a size 3 shoe was more appropriate. The child demonstrated full knee extension, so peak knee extension torque measurements (**Table 1**) were taken. His values were compared to established average norms (SD) for peak knee extension torque for typically developing 9 year-old children: 42.9(5.9) Nm[17]. After a clinical assessment consisting of a lower quarter screen, foot orthoses were fabricated with medial forefoot posting and ¼ inch heel lift. The participant was tested both with and without these custom orthoses in a women's size 7 pair of Brooks Dyads because the appropriate size shoe was unavailable.

Participant B was an 11 year-old male who was 4' 7.5'' tall, and weighed 56 pounds. He was diagnosed with DMD in 2010, at age 4, and wears a size 2 shoe. Due to the presence of bilateral knee extension lag, peak knee extension torque measurements were not taken. Fabrication of the custom orthoses led to an insert with a ⅝ inch heel lift in a Brooks Dyad shoe size 7 women's because the appropriate size shoe was unavailable. This child would also have benefited from a greater heel lift. However, due to the low height of the heel counter, a ⅝ inch heel lift was the most comfortable heel lift for the participant without the sense that his heels were being lifted out of the shoes.

Passive range of motion measurements are displayed for both participants in **Table 2**, and demonstrate that both participants had ankle equinus.

Table 1. Participant A KE Peak Torque (Nm)

Limb	Trial 1	Trial 2	Trial 3	Mean (± SD)
Left	5.1	6.8	6.9	6.3 (0.99)
Right	8.7	6.4	5.1	6.7 (1.80)

Table 2. Passive Range of Motion (°)

	Hip Extension		Knee Extension		Ankle DF (prone)		Ankle DF (sitting)	
Participant ID	Left LE	Right LE	Left LE	Right LE	Left LE	Right LE	Left LE	Right LE
A	13	15	0	0	-1	-7	-2	-2
B	20	20	-5	-9	-20	-25	-10	-10

Participants A and B scored 20/34 (**Appendix 3.1**) and 13/34 (**Appendix 3.2**), respectively, on the North Star Ambulatory Assessment without orthoses, which was conducted prior to all other balance assessments. Following orthotic fabrication, participants underwent standing and ambulatory balance testing with and without their orthoses. Static balance results are represented by **Figure 1** for Participant A, and **Figure 2** for Participant B.

Figure 1.

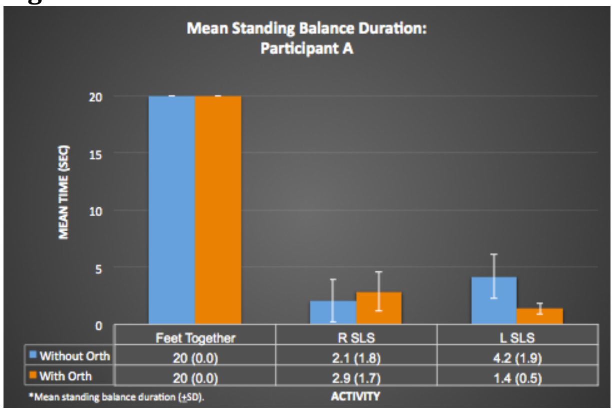
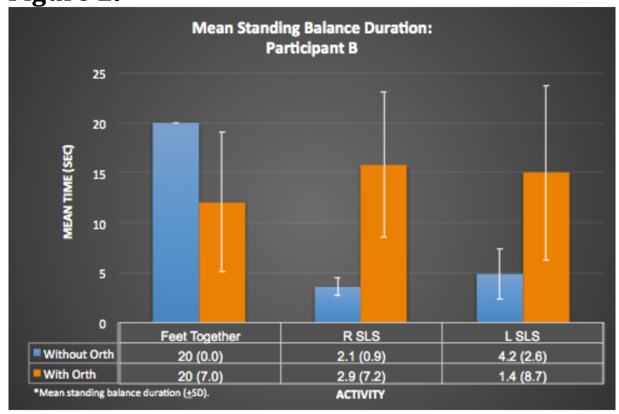


Figure 2.



Gait parameters measured by the GAITRite® software for participants A and B are represented in **Table 3** and **Table 4**, respectively. Furthermore, visual representations of their footfalls during ambulation on the GAITRite® mat is displayed for participant A in **Figure 3**, and for participant B in **Figure 4**. Images are also available to view participant B's normal stance posture and right foot strike during gait without and with orthoses (**Figure 4.1** and **Figure 4.2**, respectively).

Table 3. GAITRite® Values for Participant

PARAMETER DESCRIPTION	Without Orthoses Total/Left	Right	With Orthoses Total/Left	Right
Step Count	23.00		24.00	
Distance (cm)	974.02		1069.29	
Ambulation Time (s)	12.53		13.85	
Velocity (cm/s)	77.70		77.20	
Cadence	110.10		104.00	
Step Length Differential	1.29		0.52	
Step Length(cm)	41.68	42.97	44.83	44.32
Stride Length(cm)	83.57	84.63	92.39	88.69
Swing % of Cycle	37.50	35.70	35.60	34.90
Stance % of Cycle	62.50	64.40	64.40	65.10
Step Len Std Dev	7.86	6.61	5.02	11.68
Stride Length Std Dev	14.34	14.75	8.58	16.17
Foot Length (cm)	23.00	23.70	24.00	23.80
Foot Width (cm)	8.86	9.08	9.48	9.25

Table 4. GAITRite® Values for Participant B

PARAMETER DESCRIPTION	Without Orthoses Total/Left	Right	With Orthoses Total/Left	Right
Step Count	32.00		34.00	
Distance (cm)	1474.68		1401.54	
Ambulation Time (s)	16.56		17.55	
Velocity (cm/s)	89.10		79.90	
Cadence	115.90		116.20	
Step Length Differential	3.54		18.40	
Step Length(cm)	44.32	47.85	32.57	50.96
Stride Length(cm)	92.26	92.59	83.74	84.28
Swing % of Cycle	40.40	40.60	40.10	38.90
Stance % of Cycle	59.60	59.40	60.00	61.10
Step Len Std Dev	1.92	1.66	4.31	4.41
Stride Length Std Dev	2.31	1.93	7.70	3.62
Foot Length (cm)	10.40	9.80	20.00	12.70
Foot Width (cm)	8.93	8.89	8.12	8.81

Figure 3.

Gait Pattern without orthoses (ppt A):

*Direction of gait from left to right

Gait Pattern with orthoses (ppt A):

*Direction of gait is from right to left

Figure 4.

Gait Pattern without orthoses (ppt B):

*Direction of gait is from left to right

Gait Pattern with orthoses (ppt B):

*Direction of gait is from right to left

Figure 4.1. Stance and Right foot strike during gait without orthoses (Participant B)

Figure 4.2. Stance and Right foot strike during gait with orthoses (Participant B)

Discussion

Participant A scored a 20/34 on the North Star Ambulatory Assessment score compared to Participant B with a score of 13/34, indicating that Participant A had fewer impairments than Participant B. During this assessment, Participant A was focused and followed all instructions. Most notable in Participant B were his deficits in knee extension lag (1 deg on the left and 7 deg on the right) and ankle dorsiflexion (**Table 2**). This child was also more involved in his right lower extremity, indicated by a greater limitation in right ankle dorsiflexion in the prone position (lacking 25 deg on the right compared to lacking 20 deg on the left). These values were expected, as clinical assessment of Participant B revealed typical postural compensations and gait patterns expected in a child with DMD at his age.

Participant A presented to this study with mild physical impairments that could have diminished his balance and gait abilities. Additionally, this participant was easily distracted making it difficult to obtain reliable data. This suggests the child may not have been exerting maximal effort during testing, which is shown in his peak knee extension torque measures being well below the norms for children with DMD[18]. The screening process was completed with Participant A following a scheduled echocardiogram procedure, so fatigue may have also played a role in decreased torque measurements. Therefore, it is likely that the peak knee extension torque measures do not accurately depict this participant's true peak strength. During static and ambulatory balance assessments, the participant was easily distracted and adhering to instructions was difficult for the child. Participant A required several practice trials and encouragement to follow instructions and exert a maximal effort while extending his knee and performing balance tasks. This may contribute to him not demonstrating any statistically significant improvements in balance or gait parameters when comparing conditions with and without custom orthoses. Although he was able to complete static standing with feet together for the maximum 20 seconds timed with and without orthoses, single leg balance was difficult for him under both conditions. Fatigue may also be a potential factor in limiting Participant A's performance when completing balance tasks without orthoses due to his need for encouragement and several trials to repeat the tasks. His static single limb standing balance did not appear to be affected by the use of orthoses. However, this may also be attributed to his mild deficits, and so he may not have had much room for improvement. Similarly, the patient had difficulty focusing on walking at a baseline speed over the GAITRite® mat. Several trials of walking on the GAITRite® mat were required to obtain data that accurately depicted his typical walking pattern.

Clinical observation of both subjects has meaningful implications when evaluating the participants' movement patterns before custom orthoses fabrication and assessing the effects of the custom orthoses. Participant A's movement repertoire revealed a Gower's maneuver to be able to get up from the floor, difficulty with navigating steps without upper extremity support, and significant hip and knee extensor and hip abductor muscle weakness. While no improvements in balance or gait were apparent with the application of custom orthoses (**Table 3**), Participant B was observed to achieve better contact of each foot with the floor, ambulate with less lateral trunk sway, and improved dynamic stability. Although this participant does not yet demonstrate a significant amount of toe-walking, the increase in foot length displayed during GAITRite® walking could indicate a slight increase in base of support. When assessing Participant B during standing balance assessment with the orthoses, the child expressed an unusual sensation of a backwards weight shift due to the heel lifts and assumed a position with his bottom out (increased lumbar lordosis and flexed trunk) in order to compensate for standing with a narrow base of support. This difference in comfort in his shoes likely contributed to the decrease in bilateral stance time

[20s (SD = 0.0) without orthoses to 12.1s (SD = 7.0) with orthoses]. Even so, there was an improvement for both right and left single limb stance times [improving from 3.6s in R SLS to 15.8s (ES = 0.94), and from 4.9s to 15s in L SLS (ES = 0.97)].

For Participant B, these improvements in single limb support seemed to carry over to the GAITRite® assessment, indicated in **Figure 4**. Most prominent is the increase in left foot length measured by the GAITRite® assessment with the orthoses. This supports increased foot contact with the ground during ambulation with the orthoses, which was also clinically observed (**Figure 4.1** and **Figure 4.2**). GAITRite® data shows that participant B's left foot length nearly doubled (10.4cm to 20cm) while his right foot length during gait moderately increased (from 9.80 cm to 12.70 cm). However, the child was able to only occasionally achieve neutral landing with his right foot, which can be explained by the greater tightness in triceps surae in his right lower extremity. We noted a clinical difference in the child's gait pattern, which correlates to some of the findings on the GAITRite® data values (**Table 4**). The participant also expressed that he felt like he could walk faster with the orthoses and felt more balanced and stable during gait, despite a minimal increase in cadence (115.90 without orthoses to 116.20 with orthoses) and a decrease in velocity (89.10 cm/s without orthoses to 79.90 cm/s). Also interesting to note is the significant difference in step length differentials without orthoses (3.54) and with orthoses (18.40). We would expect the custom orthoses to improve controlled dynamic stability and specifically compensate for poor quadriceps strength, thus increasing the child's step length. However, participant B demonstrated a decrease in left step length (44.32 cm without orthoses to 32.57 cm with orthoses) and a slight increase in right step length (47.85 cm without orthoses to 50.96 cm with orthoses).

Several limitations in this study are apparent that likely affected the outcomes. Participant A was easily distracted, and it was unclear whether or not he was accurately portraying his normal balance and gait abilities throughout testing. Furthermore, participant A ambulated with toe touch at initial contact of the stance phase, but toe touch was quickly followed by heel contact with the ground, mitigating any balance issues that may be caused by decreased foot length and base of support. Screening of participant A was performed after an echocardiogram procedure on a day prior to the full balance and gait analysis. Thus, the timing of peak knee extensor torque measurement could have been a source of fatigue, which would account for the low torque values found. Participant B fatigued quickly, and required frequent rest breaks between trials. This child also presented to the study following a medical appointment with his pediatrician and reported some fatigue during testing under his second condition (with orthoses). Further, the team felt the child would benefit from additional heel lift, but was unable to provide this due to the restrictions in the height of the heel counter. Both participants presented to the clinic wearing shoes that were not ideal for using the provided orthotic inserts. The team held a discussion with participant A's family, to suggest purchasing size 3 shoes, so the orthoses could better serve the child while wearing a more appropriate shoe size. Participant B would also have benefited from different shoes with a higher heel counter (i.e., basketball shoes), as it was decided that he could benefit from another ⅛ to ¼ inch of heel lift that could not currently fit in his pair of shoes. His mother was skeptical of buying new athletic shoes, as she reported that they are typically too heavy and cause Participant B to fall more often; however, after seeing Participant B successfully walk in the Brooks Dyads, she reported that they would go search for some light basketball shoes that would allow for the additional heel lift.

At 1-month, a follow-up phone call will take place to interview the parents of each participant and provide an update on how the children have been doing with their custom orthoses.

In conclusion, the results of this study further support the anecdotal assessment that heel lifts may be a plausible intervention for ambulatory boys with DMD. As discovered in the literature review, heel lifts have shown effectiveness in treatment for individuals with limited ankle dorsiflexion range of motion[19], as well as potentially having a protective effect for the triceps surae and rectus femoris musculature[20,21]. In future studies, standardized procedures should be used to normalize GAITRite® trials between subjects to account for rest breaks and prevent fatigue or overuse. Specifically, pre-determined protocols for the number of practice trials, rest break times, and the number of passes made on the GAITRite® mat for data collection purposes may help reduce some variability between subjects. The time of day for data collection may also be an important consideration for avoiding fatigue or poor concentration. Follow-up with assessment in a motion analysis lab may help improve understanding of how movement strategies are impacted with these customized orthoses.

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Appendices

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Appendix 1. Recruitment flyer

Can custom shoe inserts make walking easier for boys with Duchenne muscular dystrophy?

We are conducting a small pilot research study of the effects of custom shoe inserts (insoles, heel lifts) on balance and walking in **boys with Duchenne muscular dystrophy who are able to walk without help. Study participants should have significant leg weakness that makes walking and other activities (such as getting up from the floor or climbing stairs) difficult for them.**

- **How much time is required?** One 2.5 hour session at University Physical and Occupational Therapy in Hillsborough, NC
- **What will study participants be asked to do?** Be evaluated and fitted with custom inserts, and complete balance and walking tests while wearing the inserts and while wearing shoes with no inserts

- **Participants will receive a pair of custom inserts for free.**

If interested, please contact Vicki Mercer at 919-843-8642 or vmercer@med.unc.edu. This study is being conducted by The University of North Carolina at Chapel Hill Division of Physical Therapy.

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Appendix 2. Screening script

Project: Immediate effects of orthoses with heel lifts on gait and balance in children with DMD
Date: _________
Screening ID Number: SCR___

Screening Questionnaire

(For "Y" or "N" questions, circle the response. If there is a box next to the circled response, place a check mark in the box. All boxes must be checked in order for the child to be eligible for the study.)

Age: ____ Date of birth: _____ Year of diagnosis: ______________

Height: _______ Weight: _______ Shoe size (include width if known):______________

What testing was done to establish your child's diagnosis of Duchenne muscular dystrophy?

Does your child speak English? Y € N

Does your child have any difficulty following instructions? Y N €
If yes, please explain:

Is your child able to walk at least 100' without help and without an assistive device? Y € N

Has your child ever been diagnosed with a neurological, neuromuscular, or cardiopulmonary disorder other than Duchenne muscular dystrophy? Y N €
If yes, please describe:

Does your child have weakness in his legs that makes it difficult for him to get up from the floor or climb stairs? Y € N

Does your child currently have any injury to or problems with his legs or feet that might make it difficult for him to use shoe inserts? Y N €
If yes, please describe:

If child is eligible based on responses above:

Does the child ambulate with forefoot strike bilaterally (as opposed to heel strike)? Y € N

Knee extension lag? If present, measure on left ______ ° and right ______°. If not present, continue:

Distance from knee joint center to center of head of handheld dynamometer ______ cm (= ______ m)

Muscle force (Newtons) for left knee extensors ________ and right knee extensors ________

Isometric muscle torque on left ________ Nm and right ________ Nm

Mean (SD) Knee Extensor Torque* in Boys

Age	5	6	7	8	9	10	11	12	13	14	15
Mean	21.0	26.0	30.2	45.4	42.9	61.4	63.3	79.3	82.5	110.4	122.1
SD	(5.8)	(4.0)	(8.5)	(12.1)	(5.9)	(14.9)	(17.0)	(12.7)	(18.3)	(23.2)	(18.6)

*Newton-meters

North Star Ambulatory Assessment

We have attempted to give clear explanations of the methods employed to achieve motor goals, but it is not possible to be exhaustive in the descriptions, particularly of modifications to activity. Whilst DMD children may generally present with recognizable adaptations to activity due to the underlying progressive muscular weakness, they may modify their activity to achieve functional goals in slightly differing ways. Generally, activities are graded in the following manner:

2 – 'Normal' – no obvious modification of activity
1 - Modified method but achieves goal independent of physical assistance from another
0 - Unable to achieve independently

Gowers' Manoeuvre:

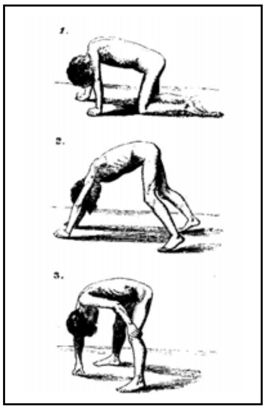

Figure: Gowers' Manoeuvre (from W.R. Gowers' *Pseudohypertrophic muscular paralysis*, 1879)

Definition of Gowers' manoeuvre:
The child turns towards the floor (generally into a four-point kneeling position) to place hands on the floor to assist rising, walks hands back in towards him then uses arms to 'climb' up legs to achieve upright standing. A wide base of support is often assumed through the phases of rising from the floor.

Stair Climb
As it is not possible to ensure standardisation, or availability, of flights of stairs, we are asking that a box step (approximately 15cm high) is used to assess single step climb and descend. A plinth or other immovable object may need to be available to provide support.

The following two pages give test details and instructions for the patient and a scoring sheet with details for grading. They should be used in conjunction. Please familiarize yourself with the test detail before starting to evaluate patients.

The **North Star Ambulatory Assessment** has been developed by the Physiotherapy Assessment and Evaluation Group of the North Star Clinical Network.
The North Star Project is supported by
Muscular Dystrophy Campaign

For further information contact Elaine Scott, North Star Project Coordinator
elaines@muscular-dystrophy.org

Test Detail and Instructions to Patient

Activity	Instructions to patient	Start position/test detail	Comments
1. Stand	Can you stand up tall for me for as long as you can and as still as you can	Feet should be close together and heels on the ground if possible. Arms by sides. NO shoes should be worn.	Best done on the floor rather than on a mat. Whichever is chosen maintain consistency through repeated testing sessions. Minimum count of 3 seconds to score 2.
2. Walk	Can you walk from A to B (state to and where from) for me.	Walk without shoes/socks on. Should be enough of a distance to observe 'normal gait' for that subject	A value judgement needs to be made in scoring – if the patient generally toe walks but occasionally gets heels flat, or can on request but doesn't usually, they should score 1
3. Stand up from chair	Stand up from the chair keeping your arms folded if you can	Starting position 90º hips and knees, feet on floor/supported on a box step.	A size-appropriate chair or height adjustable plinth should be used. Arms should be kept crossed throughout the activity to score 2.
4. Stand on one leg - Right	Can you stand on your right leg for as long as you can?	Minimum count of 3 seconds to score 2. NO shoes should be worn.	Best done on the floor rather than on a mat. Whichever is chosen maintain consistency through repeated testing sessions.
5. Stand on one leg - Left	Can you stand on your left leg for as long as you can?	Minimum count of 3 seconds to score 2. NO shoes should be worn.	Best done on the floor rather than on a mat. Whichever is chosen maintain consistency through repeated testing sessions.
6. Climb box step - right	Can you step onto the top of the box using your right leg first?	Stands facing the box step. Step should be approximately 15cm high	Support may be provided by the use of a height adjustable plinth, or, if not available a 'neutral' hand from the therapist.
7. Climb box step - left	Can you step onto the top of the box using your left leg first?	Stands facing the box step. Step should be approximately 15cm high	Support may be provided by the use of a height adjustable plinth, or, if not available a 'neutral' hand from the therapist.
8. Descend box step - Right	Can you step down from the box using your right leg first?	Stands on top of the box step facing forwards. Step should be approximately 15cm high	Support may be provided by the use of a height adjustable plinth, or, if not available a 'neutral' hand from the therapist.
9. Descend box step - Left	Can you step down from the box using your left leg first?	Stands on top of the box step facing forwards. Step should be approximately 15cm high	Support may be provided by the use of a height adjustable plinth, or, if not available a 'neutral' hand from the therapist.
10. Gets to sitting	Can you get from lying to sitting?	Starting position supine on a mat. No pillow should be used under head	If patient turns into prone or towards the floor to work their way into sitting 1 should be scored
11. Rise from floor	Get up from the floor using as little support as possible and as fast as you can (from supine)	Starting position supine with arms by sides, legs straight. No pillow to be used	Activity should be attempted without use of furniture in the first instance. Do not note time if a chair has to be used.
12. Lifts head	Lift your head to look at your toes keeping your arms folded	Supine on a mat. No pillow should be used.	Ask patient to keep arms crossed over chest during the activity to avoid self-assist. Also ask to look at toes to ensure neck is flexed – should be a chin to chest manoeuvre.
13. Stands on heels	Can you stand on your heels?	Standing on the floor. No shoes to be worn.	Watch for inversion. If substantial inversion but forefeet are still lifted – score 1. If only inversion with lateral border of foot still on the ground score 0.
14. Jump	How high can you jump?	Standing on the floor, feet fairly close together.	Want height, not forward movement. Small amount of forward movement acceptable
15. Hop right leg	Can you hop on your right leg?	Starting position standing on floor on right leg. No shoes should be worn.	Needs obvious floor clearance to score 2
16. Hop left leg	Can you hop on your left leg?	Starting position standing on floor on right leg. No shoes should be worn.	Needs obvious floor clearance to score 2
17. Run (10m)	Run as fast you can to……(give point)	A straight 10m walkway should be clearly marked in a quiet department or corridor. A stopwatch should be used to time the walk. Be consistent as to whether shoes are worn or not. Ensure safety of patient. They should self select speed after being asked to go 'as fast as they can'.	'Duchenne jog' - not a true run (there probably IS a double support phase), but more than a walk. Typically characterized by excessive use of arms, trunk rotation, substantial 'waddle'. No real 'push-off'

North Star Ambulatory Assessment – Score Sheet

Activity	2	1	0	Comments
1. Stand	Stands upright, still and symmetrically, without compensation (with heels flat and legs in neutral) for minimum count of 3 seconds	Stands still but with some degree of compensation (e.g. on toes or with legs abducted or with bottom stuck out) for minimum count of 3 seconds	Cannot stand still or independently, needs support (even minimal)	
2. Walk	Walks with heel-toe or flat-footed gait pattern	Persistent or habitual toe walker, unable to heel-toe consistently	Loss of independent ambulation – may use KAFOs or walk short distances with assistance	
3. Stand up from chair	Keeping arms folded Starting position 90º hips and knees, feet on floor/supported on a box step.	With help from thighs or push on chair or prone turn	Unable	
4. Stand on one leg - right	Able to stand in a relaxed manner (no fixation) for count of 3 seconds	Stands but either momentarily or needs a lot of fixation e.g. by knees tightly adducted or other trick	Unable	
5. Stand on one leg - left	Able to stand in a relaxed manner (no fixation) for count of 3 seconds	Stands but either momentarily or needs a lot of fixation e.g. by knees tightly adducted or other trick	Unable	
6. Climb box step - right	Faces step – no support needed	Goes up sideways or needs support	Unable	
7. Climb box step - left	Faces step – no support needed	Goes up sideways or needs support	Unable	
8. Descend box step - right	Faces forward, climbs down controlling weight bearing leg. No support needed	Sideways, skips down or needs support	Unable	
9. Descend box step -left	Faces forward, climbs down controlling weight bearing leg. No support needed	Sideways, skips down or needs support	Unable	
10. Gets to sitting	Starts in supine – may use one hand to assist	Self assistance e.g. – pulls on legs or uses head-on-hands or head flexed to floor	Unable	
11. Rise from floor	From supine – no evidence of Gowers' manoeuvre*	Gowers' evident	(a) NEEDS to use external support object e.g. chair OR (b) Unable	Time (00.0s)…………………
12. Lifts head	In supine, head must be lifted in mid-line. Chin moves towards chest	Head is lifted but through side flexion or with no neck flexion	Unable	
13. Stands on heels	Both feet at the same time, clearly standing on heels only (acceptable to move a few steps to keep balance) for count of 3	Flexes hip and only raises forefoot	Unable	
14. Jump	Both feet at the same time, clear the ground simultaneously	One foot after the other (skip)	Unable	
15. Hop right leg	Clears forefoot and heel off floor	Able bend knee and raise heel, no floor clearance	Unable	
16. Hop left leg	Clears forefoot and heel off floor	Able bend knee and raise heel, no floor clearance	Unable	
17. Run (10m)	Both feet off the ground (no double stance phase during running)	'Duchenne jog'	Walk	Time (00.0s)…………………
				TOTAL= /34

* See definition page 1

Appendix 3.1. Participant A North Star Score Sheet

North Star Ambulatory Assessment – Score Sheet

Activity	2	1	0	Comments
1. Stand	Stands upright, still and symmetrically, without compensation (with heels flat and legs in neutral) for minimum count of 3 seconds	Stands still but with some degree of compensation (e.g. on toes or with legs abducted or with bottom stuck out) for minimum count of 3 seconds	Cannot stand still or independently, needs support (even minimal)	2
2. Walk	Walks with heel-toe or flat-footed gait pattern	Persistent or habitual toe walker, unable to heel-toe consistently	Loss of independent ambulation – may use KAFOs or walk short distances with assistance	1
3. Stand up from chair	Keeping arms folded Starting position 90º hips and knees, feet on floor/supported on a box step.	With help from thighs or push on chair or prone turn	Unable	1
4. Stand on one leg - right	Able to stand in a relaxed manner (no fixation) for count of 3 seconds	Stands but either momentarily or needs a lot of fixation e.g. by knees tightly adducted or other trick	Unable	2
5. Stand on one leg - left	Able to stand in a relaxed manner (no fixation) for count of 3 seconds	Stands but either momentarily or needs a lot of fixation e.g. by knees tightly adducted or other trick	Unable	2
6. Climb box step - right	Faces step – no support needed	Goes up sideways or needs support	Unable	2
7. Climb box step - left	Faces step – no support needed	Goes up sideways or needs support	Unable	1
8. Descend box step - right	Faces forward, climbs down controlling weight bearing leg. No support needed	Sideways, skips down or needs support	Unable	2
9. Descend box step -left	Faces forward, climbs down controlling weight bearing leg. No support needed	Sideways, skips down or needs support	Unable	1
10. Gets to sitting	Starts in supine – may use one hand to assist	Self assistance e.g. – pulls on legs or uses head-on-hands or head flexed to floor	Unable	2
11. Rise from floor	From supine – no evidence of Gowers' manoeuvre*	Gowers' evident	(a) NEEDS to use external support object e.g. chair OR (b) Unable	Time (00.0s)……1……………
12. Lifts head	In supine, head must be lifted in mid-line. Chin moves towards chest	Head is lifted but through side flexion or with no neck flexion	Unable	2
13. Stands on heels	Both feet at the same time, clearly standing on heels only (acceptable to move a few steps to keep balance) for count of 3	Flexes hip and only raises forefoot	Unable	1
14. Jump	Both feet at the same time, clear the ground simultaneously	One foot after the other (skip)	Unable	0
15. Hop right leg	Clears forefoot and heel off floor	Able bend knee and raise heel, no floor clearance	Unable	0
16. Hop left leg	Clears forefoot and heel off floor	Able bend knee and raise heel, no floor clearance	Unable	0
17. Run (10m)	Both feet off the ground (no double stance phase during running)	'Duchenne jog'	Walk	Time (00.0s)…1;.4.37.sec..
				TOTAL= 20**/34**

* See definition page 1

North Star Ambulatory Assessment – Score Sheet

Activity	2	1	0	Comments
1. Stand	Stands upright, still and symmetrically, without compensation (with heels flat and legs in neutral) for minimum count of 3 seconds	Stands still but with some degree of compensation (e.g. on toes or with legs abducted or with bottom stuck out) for minimum count of 3 seconds	Cannot stand still or independently, needs support (even minimal)	2
2. Walk	Walks with heel-toe or flat-footed gait pattern	Persistent or habitual toe walker, unable to heel-toe consistently	Loss of independent ambulation – may use KAFOs or walk short distances with assistance	1
3. Stand up from chair	Keeping arms folded Starting position 90º hips and knees, feet on floor/supported on a box step.	With help from thighs or push on chair or prone turn	Unable	1
4. Stand on one leg - right	Able to stand in a relaxed manner (no fixation) for count of 3 seconds	Stands but either momentarily or needs a lot of fixation e.g. by knees tightly adducted or other trick	Unable	1
5. Stand on one leg - left	Able to stand in a relaxed manner (no fixation) for count of 3 seconds	Stands but either momentarily or needs a lot of fixation e.g. by knees tightly adducted or other trick	Unable	1
6. Climb box step - right	Faces step – no support needed	Goes up sideways or needs support	Unable	1
7. Climb box step - left	Faces step – no support needed	Goes up sideways or needs support	Unable	1
8. Descend box step - right	Faces forward, climbs down controlling weight bearing leg. No support needed	Sideways, skips down or needs support	Unable	1
9. Descend box step -left	Faces forward, climbs down controlling weight bearing leg. No support needed	Sideways, skips down or needs support	Unable	1
10. Gets to sitting	Starts in supine – may use one hand to assist	Self assistance e.g. – pulls on legs or uses head-on-hands or head flexed to floor	Unable	1
11. Rise from floor	From supine – no evidence of Gowers' manoeuvre*	Gowers' evident	(a) NEEDS to use external support object e.g. chair OR (b) Unable	Time (00.0s)………………… 1
12. Lifts head	In supine, head must be lifted in mid-line. Chin moves towards chest	Head is lifted but through side flexion or with no neck flexion	Unable	0
13. Stands on heels	Both feet at the same time, clearly standing on heels only (acceptable to move a few steps to keep balance) for count of 3	Flexes hip and only raises forefoot	Unable	0
14. Jump	Both feet at the same time, clear the ground simultaneously	One foot after the other (skip)	Unable	0
15. Hop right leg	Clears forefoot and heel off floor	Able bend knee and raise heel, no floor clearance	Unable	0
16. Hop left leg	Clears forefoot and heel off floor	Able bend knee and raise heel, no floor clearance	Unable	0
17. Run (10m)	Both feet off the ground (no double stance phase during running)	'Duchenne jog'	Walk	Time (00.0s)……1……………
				TOTAL= 13**/34**

* See definition page 1

Appendix 4. Follow-up Questionnaire

Follow-Up Interview

How is you child doing? Is he still able to walk without help?

Is your child still wearing the shoe inserts?

If so, about how many hours per day on average?

If not, why not?

On a scale of 1-10, with 1 being not helpful at all and 10 being very helpful, how helpful were/are the shoe inserts in improving your child's balance?

On a scale of 1-10, with 1 being not helpful at all and 10 being very helpful, how helpful were/are the shoe inserts in improving your child's walking?

What are/were some things about the shoe inserts that you and/or your child like(d)?

What are/were some things about the shoe inserts that you and/or your child don't (didn't) like?