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Ankle bracing practices in ambulatory, corticosteroid-naive boys with Duchenne muscular dystrophy

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Abstract

Introduction

Loss of ambulation in Duchenne muscular dystrophy presages scoliosis, respiratory failure, and death. Strategies to maintain ankle range of motion are employed, but little evidence exists to support these approaches and limited information is available concerning current practice.

Methods

In this study we assessed baseline bracing data from 187 boys participating in a multicenter, international clinical trial.

Results

Ankle-foot orthoses (AFOs) were recommended for 54% of the boys, with nighttime static AFOs and nighttime dynamic AFOs utilized in 94% and 6% of these boys, respectively. Daytime static AFOs were recommended for 3 boys. Compliance with bracing recommendations was 54% for nighttime static braces and 67% for nighttime dynamic braces.

Discussion

The basis for the variation in recommended AFO use is unknown and requires further study. Long-term follow-up of boys may permit assessment of the effects of AFO use.

Abbreviations

ADF
ankle dorsiflexion
AFO
ankle-foot orthoses
CE
clinical evaluator
CRF
case report form
DMD
Duchenne muscular dystrophy
FOR-DMD trial
Finding the Optimal Steroid Regimen in Duchenne Muscular Dystrophy trial
FVC
forced vital capacity
NSAA
North Star Ambulatory Assessment
PT
physical therapist
ROM
range of motion

1 INTRODUCTION

In Duchenne muscular dystrophy (DMD), loss of ankle range of motion (ROM), specifically dorsiflexion, is well documented.[1, 2] As weakness progresses, extensibility of muscle tissue is lost due to fibrosis and tissue damage. As strength declines, boys routinely use postural compensations to maintain the ability to stand and walk, including widening the base of support, avoidance of hip-extension excursion, retraction of the shoulder girdle, and accentuation of lumbar lordosis, all of which shift the center of mass forward and reduce the strain on weakened muscle groups.[3] This forward weight shift reinforces toe walking, further shortening the gastrocnemius/soleus complex with reduced excursion into ankle dorsiflexion (ADF) in the gait cycle. These modifications result in a loss of efficient, safe, propulsive gait. Infrequent excursion into dorsiflexion in the gait cycle, coupled with the cellular changes in the muscles themselves, cause muscle stiffness and gradual loss of ankle ROM.

Maintenance of functional ankle ROM remains important for standing, ambulation, and functional mobility. Early recognition of the importance of ankle ROM is assumed to be a key to a functional program and is considered best practice in physical therapy.[1, 4, 5] To achieve prolonged positional stretching of the Achilles tendons, calves, and ankle soft tissues, a variety of positional stretching devices may be applied during the day or overnight. Options include bi-valved casts, static ankle-foot orthoses (AFOs) (fixed at one angle, typically neutral or 0°), and dynamic AFOs, which include a hinged ankle and one of several different types mechanisms to produce low load stress into dorsiflexion. These braces are designed to be worn for long periods, typically overnight, and are not for use while walking as they provide continual low load-stretching stress, which would interfere with the ankle actions during walking.

Currently, there is no reliable evidence indicating which of many different types of interventions for ankle ROM management is most effective or best tolerated by patients and families, nor whether preserving ADF ROM is associated with or supports

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prolonged ambulation. No evidence yet exists to indicate at what time, age, or joint flexibility level intervention should be initiated. The Finding the Optimal Steroid Regimen in Duchenne Muscular Dystrophy (FOR-DMD) trial gathered data on a large group of boys with DMD that included information on ankle passive range of dorsiflexion motion, the type of AFOs recommended, and the recommended and actual wearing times. We examined baseline data from the FOR-DMD trial to describe current international practices of prescribing different AFOs in a cohort of DMD patients from five countries. We expected to find differences between countries with regard to AFO prescription, given different health-care systems, and expected to find similar compliance across different styles of bracing given that the recommendations were being made by perceived experts in DMD care.

2 METHODS

The FOR-DMD trial is an international, multicenter, randomized, double-blind, parallel-group trial comparing 3-year outcomes for the three most commonly used corticosteroid regimens. The trial is being performed at 32 sites in five countries (United States, Canada, United Kingdom, Italy, and Germany) in genetically confirmed DMD boys from age 4 to 7 years 11 months (at randomization), although two boys age 8.1 years were ultimately enrolled. Boys were required to be steroid-naive and able to walk at least 6 minutes to enter the study. Details of this study have been published.[6] Physical therapy assessments include measurements of passive ROM, muscle extensibility, posture, strength, and function using the North Star Ambulatory Assessment (NSAA) and timed function studies.[7, 8] The study was approved by the local institutional review board/ethics committee at each participating institution. Written informed consent was obtained from the parents/legal guardians of the study participants.

The clinical evaluators (CEs) at the different sites underwent hands-on training with the lead physical therapists (PTs) of the FOR-DMD trial. Reliability of the assessments was verified via review by the lead PTs of videotapes of the assessments of the first two patients enrolled at each site. Feedback for consistency and accuracy was provided back to the CE by phone call or meeting.

In this study, because interest was in the association between gastrocnemius shortening and ankle dorsiflexion, measurement of ankle dorsiflexion was undertaken in the supine position with the knee in full extension. The calcaneus was held in neutral alignment while pressure was applied over the midsection of the foot to dorsiflex the ankle as much as possible, preventing inversion. The axis of the goniometer was over the lateral malleolus, with the stationary arm aligned with the fibular head along the lateral aspect of the lower leg. The “moving arm” was held parallel to the lateral aspect of the fifth metatarsal, and aligned with the posterior third of the foot to ensure the gastrocnemius range was being monitored and not that of the plantar structures of the foot. The range of dorsiflexion past plantargrade was noted as +X° and range lacking from plantargrade as −X°. Normal range is 20° dorsiflexion to 50° plantarflexion.[9] The CE used an appropriately sized goniometer to measure ankle ROM to the nearest degree. Passive range was measured so that the subject needed to relax, whereas the CE ranged the joint. Measurements were taken with a “moderate” degree of stretch applied and with the child trying to dorsiflex the foot at the same time. The child was asked to count to ten while measuring the range.

Data from the trial regarding the use of brace interventions for ankle range of motion specifically were entered by the CE at each site under the following headings: Static AFO; Contracture Correction Device; Serial Casting; and Stretching Exercises. With the input of several lead PTs in each participating country, descriptions of the braces they documented under each heading were gathered. “Static AFO” was universally applied to mean a rigid plastic AFO with nonelastic straps across the proximal tibia and ankle mortise, possibly also across the forefoot. These are generally custom molded to each patient and are set at neutral ankle dorsiflexion (0°) with a neutral hindfoot (0° of calcaneal varus/valgus) and allow no motion of the ankle joint. Under “Contracture Correction Device,” most PTs documented any type of jointed or hinged brace that provided some low load stress into ADF by either nonelastic straps or a mechanical spring-loaded hinge. These are generally designed to provide and allow gradual motion into greater degrees of ADF over several hours of use as the soft tissue relaxes and extends in response to the loading stress. In Canada, a process of serial splinting was occasionally used whereby trained occupational therapists mold new static splints periodically in response to increasing range of motion. By report of one of the Canadian PTs, this process was not documented on the case report forms (CRFs) as it did not fall clearly into any category. As this study focused on the brace interventions, data on other stretching strategies are not included in the analyses.

In addition to the specific style of brace recommended by the CE, the prescribed frequency and duration and the reported compliance (documented as hours used) were collected. Compliance was determined through documentation of the parental reports on actual use.

Pairwise comparisons among groups defined by type of AFO used were compared with respect to age and ADF ROM, using Wilcoxon rank sum tests. All statistical analyses were performed using SAS version 9.4 (SAS Institute, Cary, North Carolina). A significance level of 5% was used for hypothesis testing.

3 RESULTS

There were 196 boys randomized in the FOR-DMD trial. Baseline data on bracing were available in 187 (95.4%) of these boys (Table 1). Fifty-four percent of boys received a recommendation for bracing of any kind (ie, 101 of 187), mostly for nighttime static AFOs (N = 95). The frequency of recommendation of nighttime use of a static AFO varied across countries as follows: 46 of 78 (59%) in the United States; 11 of 22 (50%) in Italy; 28 of 57 (49%) in the United Kingdom; 5 of 12 (42%) in Canada; and 5 of 18 (28%) in Germany. Fifty-one occurrences of actual use of a static AFO at night were documented; therefore, 44 families (46%) did not comply with this recommendation.

Table 1. Characteristics of enrolled participants

Category	Mean (SD) or number (%)
Age (years)	5.9 (1.0)
Degrees of ROM of ankle dorsiflexion	
Left	5.6 (9.1)
Right	5.7 (9.4)
Bracing recommendation	
None	86 (46.0%)

Category	Mean (SD) or number (%)
Static daytime	2 (1.1%)
Static nighttime	92 (49.2%)
Static daytime and nighttime	1 (0.5%)
Static and dynamic nighttime	2 (1.1%)
Dynamic nighttime	4 (2.1%)
Bracing use	
Static daytime (n = 4 *)	

Abbreviations: ROM, range of motion; SD, standard deviation.

* Daytime static ankle-foot orthoses were used by 5 boys; data on frequency/hours of use were missing for 1 boy.

† Nighttime static AFOs were used by 51 boys; data on frequency of use were missing for 3 boys and data on hours of use were missing for 5 boys.

Daytime use of static AFOs was rarely recommended at any site. It was recommended for three boys (1.6%; 1 each in the United States, Canada, and Italy), whereas it was documented as used by five boys (2.7%; 4 in the United States and 1 in Canada).

The use of "contracture correction devices" or dynamic AFOs, was recommended for two boys (10%) in Germany and four boys (5%) in the United States. Neither of the two German boys complied with the recommendation, whereas all four of boys in the United States reported compliance. The remaining countries did not recommend nor use dynamic AFOs. No one documented the use of serial casting.

No statistically significant differences were noted between type of AFO employed and age (Figure 1) or ADF ROM (Figure 2). The small sample sizes for all types of AFO used, other than nighttime use of static AFOs, precluded meaningful comparisons. We noted that three of the four boys using dynamic AFOs were under the age of 5.5 years, whereas the ages of boys using static AFOs tended to be somewhat higher (Figure 1). A smaller ADF ROM tended to be associated with use of the dynamic style of brace (at or just below 0° or neutral), whereas static braces tended to be more often used with initial passive dorsiflexion greater than 0° (Figure 2), although comparisons are limited by the small sample sizes. There was little relationship between ankle ROM and age, although there were some boys who exhibited more impairment after the age of 7 years (see Figure S1 online).

Figure 1 Open in figure viewer | PowerPoint

Distribution of age by use of AFOs. Each dot indicates an individual subject. AFOs, ankle-foot orthoses

Figure 2 Open in figure viewer | PowerPoint

Distribution of ADF ROM, measured as degrees from plantargrade, by use of AFOs. Data are shown for the left (A) and right (B) ankles. ADF ROM, ankle dorsiflexion range of motion; AFOs, ankle-foot orthoses

4 DISCUSSION

The standards of care for DMD include broad recommendations for a spectrum of therapeutic interventions for ROM maintenance, including stretching, bracing, and use of equipment to achieve supported standing position. Few details have been presented, and prescription based on the published standards requires each PT to determine the "best" options.5

Bracing was used in just over 50% of the patients for prevention of ankle plantarflexion contractures in the five countries participating in the FOR-DMD trial. As all participants entered the study with the ability to walk functional distances (as measured by the 6-minute walk test), the lack of use of static AFOs during the day seems to confirm that permitting ambulatory compensations is practiced in the five countries in FOR-DMD. Wearing a static AFO during the day would limit the participant's ability to walk by preventing functional compensations for weakness or would require seated time during the day for prolonged stretching. Enforced sitting in an otherwise ambulatory boy with DMD is generally believed to be counterproductive to prolongation of the ambulatory stage of the disease.10, 11 This is the likely reason serial casting appears to no longer be used in the five countries in the study, as even fairly lightweight fiberglass casts limit ambulation in boys with progressive weakness. Serial casting would increase resistance to leg motions during walking, thereby overstressing fragile muscle tissue and possibly accelerating disease progression.12 The fact that more than half of the boys received a prescription for some kind of bracing indicates that many PTs are applying strategies of prolonged positional stretching in their practice. However, there was no uniform practice with regard to bracing prescriptions. The lack of a mechanism to verify that the brace prescription came only from the CE may also have impact on the nonuniform prescription patterns identified. This variability may be linked to several issues with regard to treating boys with DMD, such as lack of availability of certain types of braces, lack of training, reliance on other strategies to address ADF ROM, and the different theoretical paradigms.

Contrary to our hypothesis, the differences we observed are not statistically significant, possibly due to the small cohorts in each bracing category. In this study we have provided a description of the current bracing practices as observed in the FOR-DMD study and, therefore, we are unable to make strong recommendations at this time. We also cannot draw any conclusions concerning differences in compliance between the types of braces prescribed due to the small number of boys who received a prescription for the dynamic type.

Although static AFOs at night were the most frequently recommended, there appeared to be a trend toward recommending dynamic AFOs when the initial ADF ROM was more reduced. However, the conclusions one can draw are limited by the small number of boys using dynamic AFOs (n = 4). It is not clear why boys who presented with impaired ADF ROM did not receive a brace prescription. There are multiple factors involved in prescribing, choosing, and obtaining different styles of brace. These factors include availability, cost, reimbursement, ease of use, family compliance, and therapist and physician training and experience.

Prolonged positional stretching is one strategy to address ankle ROM management. Outpatient or school-based skilled physical therapy, home programs of manual or exercise-based stretching, and use of standing frames (devices that maintain a patient in static standing for a set period of time) are also used. Reporting on these strategies was included in the FOR-DMD assessments but was not included in this analysis as there was no standardization of skilled physical therapy treatments and no details on the selection and style of standing frames. Therefore, meaningful comparisons were not be possible.

Although these initial data have revealed variability in ROM intervention use in a large sample of young boys in the ambulatory stages of DMD, many more questions arisesuch as: (1) What are the main factors underlying the choice to recommend or not recommend bracing in the ambulant DMD boy? (2) Does bracing preserve ADF ROM? (3) Does preserving ADF ROM prolong ambulation? Evidence is still lacking for the effectiveness of prolonged positional stretching of the Achilles tendon and posterior calf structures in prolonging ambulation for boys with DMD. Evidence is also lacking for determination of the most efficacious interventions.

The present findings represent a starting point from which further studies of preservation of ADF ROM can proceed. As longitudinal data become available from the FOR-DMD study, we will assess the impact of different interventions on long-term ankle flexibility and walking. This information can guide best practice for PTs and physicians toward the most efficient strategies to support prolongation of ambulation. FOR-DMD is an interventional study with respect to corticosteroids, but is only observational with respect to ankle bracing, thus limiting our ability to draw conclusions regarding causal relationships involving the latter type of therapy. There may be variation due to different prescribing practices in each country, whether the care of the DMD patient was overseen by different types of physicians, and the level of independence of the PT in prescribing the intervention. Statistical methods for causal inference in observational studies may be useful in addressing these issues.[13] In addition, the fact that the boys were on one of three possible steroid regimens may have impacted the walking outcomes, which will be clarified in future longitudinal data from this study. Further studies looking at other stretching interventions and bracing would shed additional light and provide guidance on brace prescription and best practices.

PTs rely on their experience with braces to recommend styles. When prescribing for the DMD population, they would be best served by having a discussion about the child's current sleep schedule and any sleeping problems, as nighttime splinting must not interfere with sleep hygiene. In addition, if the PT is unfamiliar with dynamic brace styles and options available in their area, working with an experienced certified orthotist may prove fruitful to their practice. Our study represents a starting point for subsequent work to standardize care and institute best practices for the care of boys with DMD. The longitudinal data from the FOR-DMD study may also provide information concerning whether consistent, early use of ankle bracing impacts subsequent disability, as defined by walking measures. As we await the conclusion of the FOR-DMD trial, we recommend that PTs prescribe bracing that they believe would be best tolerated by the patient and additional stretching interventions to maintain ankle range of motion and support prolonged ambulation.

ACKNOWLEDGMENTS

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CONFLICTS OF INTEREST

The authors declare no potential conflicts of interest.

ETHICAL PUBLICATION STATEMENT

We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.

Supporting Information

Filename	Description
mus26727-sup-0001-FigureS1.docx Word 2007 document , 38.3 KB	**FIGURE S1** Associations between age and ankle dorsiflexion ROM for the left and right limbs. The color of each point indicates the type(s) of AFOs used. Abbreviations: AFOs, ankle-foot orthoses; ROM, range of motion.

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Table 1 (full view)

Full Table 1

Category	Mean (SD) or number (%)
Age (years)	5.9 (1.0)
Degrees of ROM of ankle dorsiflexion	
Left	5.6 (9.1)
Right	5.7 (9.4)
Bracing recommendation	
None	86 (46.0%)
Static daytime	2 (1.1%)
Static nighttime	92 (49.2%)
Static daytime and nighttime	1 (0.5%)
Static and dynamic nighttime	2 (1.1%)
Dynamic nighttime	4 (2.1%)
Bracing use	
Static daytime (n = 4*)	
Days/week	6.0 (1.2)
Hours/week	17.8 (21.5)
Static nighttime	
Days/week (n = 48†)	6.4 (1.4)
Hours/week (n = 46†)	42.4 (28.0)
Dynamic nighttime (n = 4)	
Days/week	7.0 (0.0)
Hours/week	45.3 (17.2)

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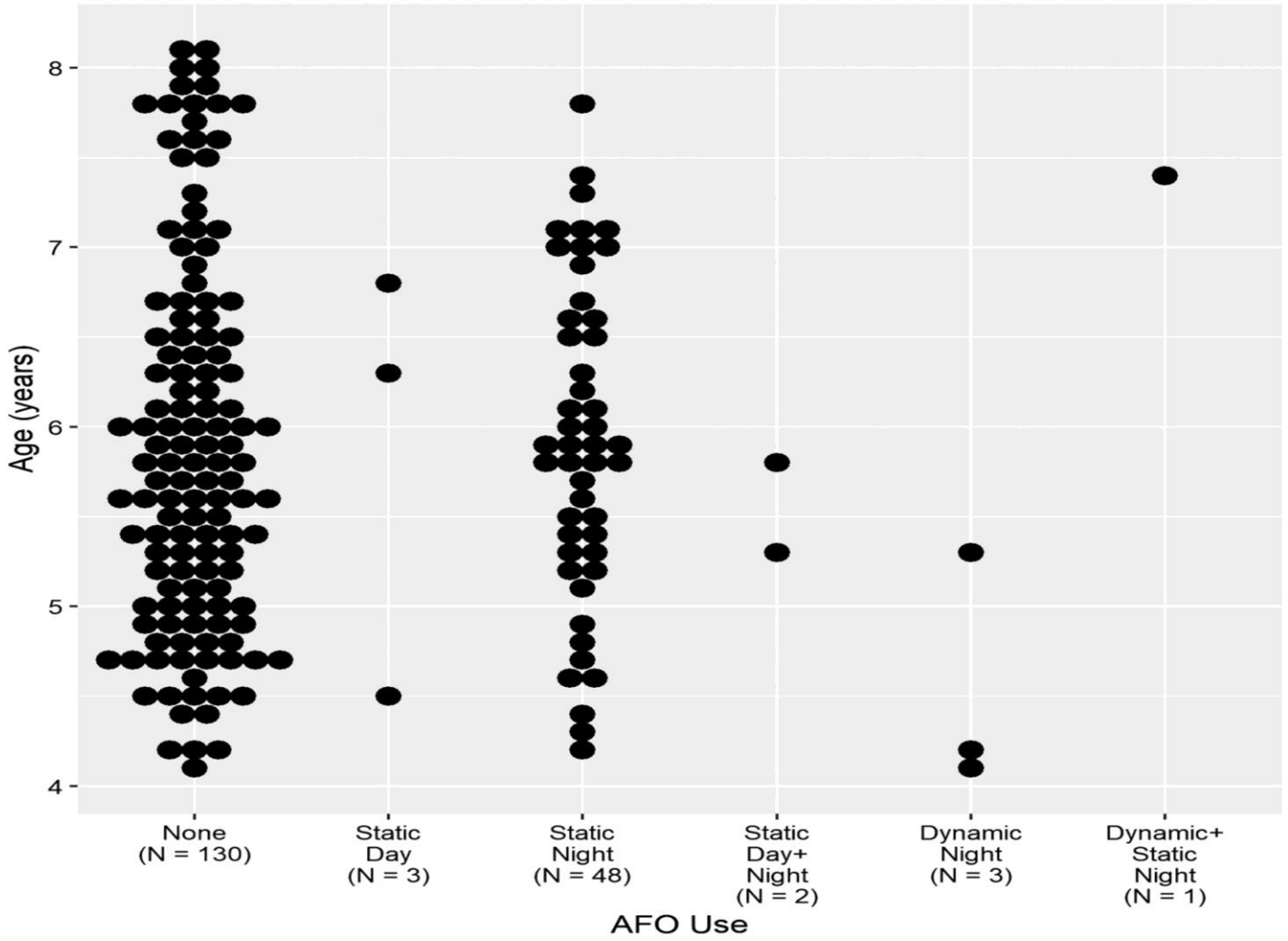
Ankle bracing practices in ambulatory, corticosteroid-naive boys with Duchenne muscular dystrophy

Figure 1



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Ankle bracing practices in ambulatory, corticosteroid-naive boys with Duchenne muscular dystrophy

Figure 2

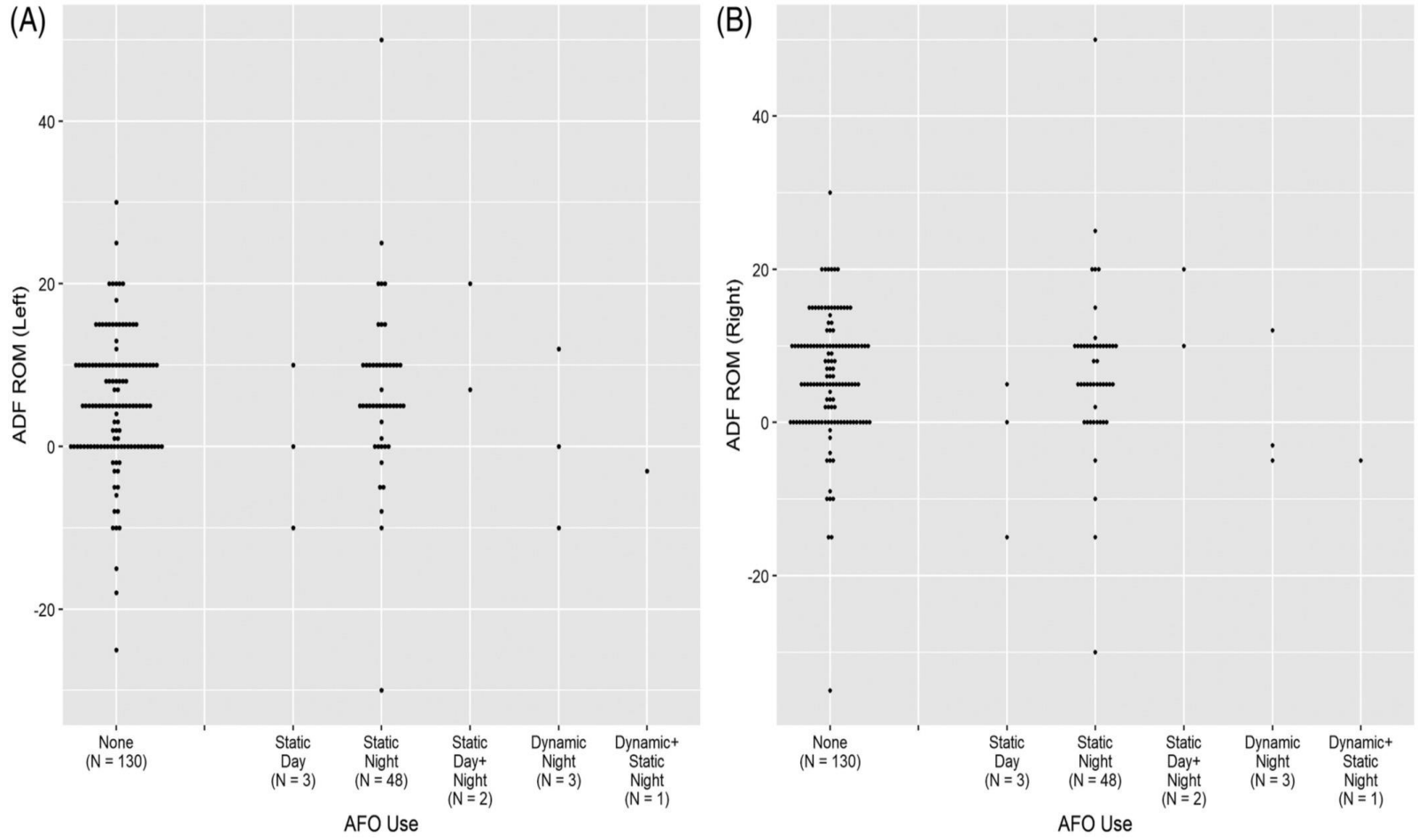

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Figure S1(LEFT). Associations between age and ankle dorsiflexion range of motion (ROM) for the left and right limbs. The color of each point indicates the type(s) of AFO used.

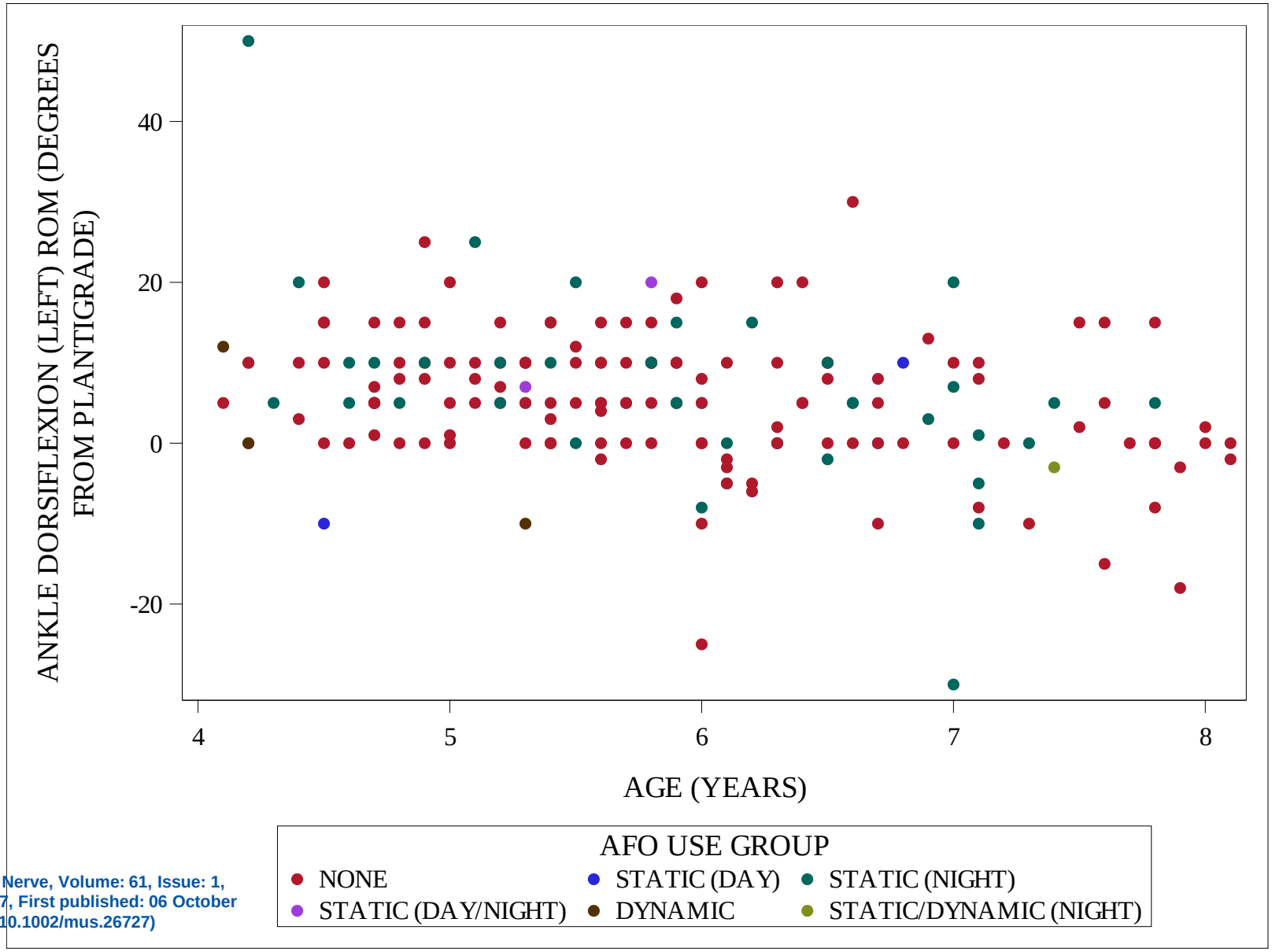

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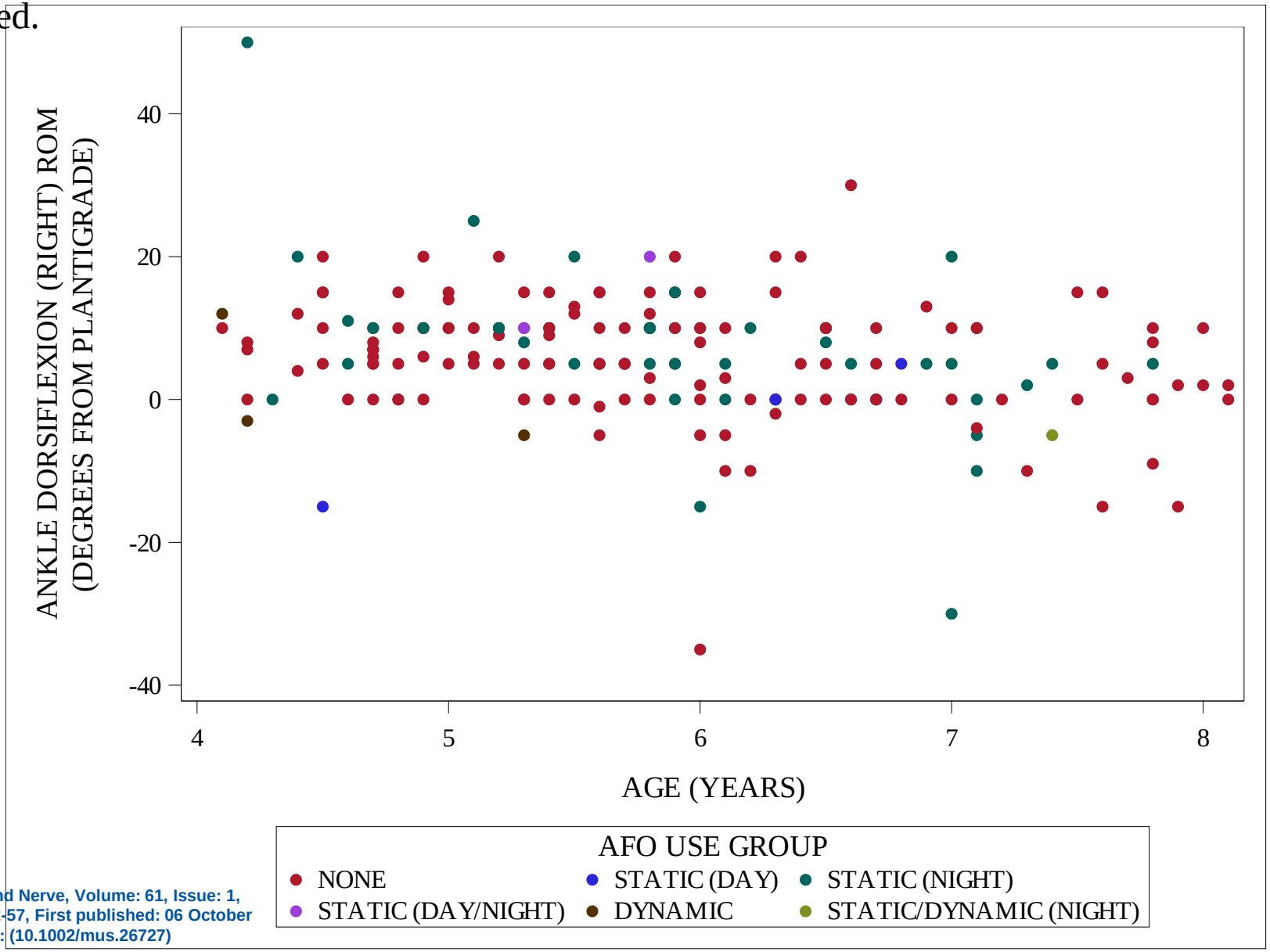
Figure S1(RIGHT). Associations between age and ankle dorsiflexion range of motion (ROM) for the left and right limbs. The color of each point indicates the type(s) of AFO used.



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