

Effect of using orthoses on prolonging ambulation in patients with Duchenne Muscular Dystrophy: review of literature

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ABSTRACT

The walking ability of patients with Duchenne muscular dystrophy gradually decreases due to advancing weakness and muscle contracture. Lower limb orthoses are often prescribed in an attempt to prolong ambulation in these patients. **Objective:** To perform a literature review in order to verify the relationship between using orthoses and prolonging ambulation. **Method:** A literature review was performed in the PUBMED, PEDRO, and SCIELO databases with the keywords orthoses, bracing, gait, gait loss, ambulation, and Duchenne muscular dystrophy. **Results:** In 14 selected articles the prescription of knee-ankle-foot orthoses (KAFO) (also called long orthoses) and ankle-foot orthoses (AFO) was identified, always associated with another therapeutic intervention. Most studies have reported that the use of such a device prolongs ambulation. **Conclusion:** The use of orthoses, regardless of type, prolongs ambulation, because it delays the progress of muscle contracture. Thus, the early use of AFO is recommended in order to minimize the functional impairment characteristic of the disease.

Keywords: Muscular Dystrophy, Duchenne, Orthotic Devices, Gait

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INTRODUCTION

In Duchenne Muscular Dystrophy (DMD), the loss of ambulation occurs at around nine years of age and it is the consequence of progressive muscle weakness and advances in muscle contracture that afflict the lower limbs.^{1,2} Despite the progression of the disease being inevitable, an important goal in the treatment of these patients is to prolong their ability to ambulate.³ It has been suggested that maintaining their gait minimizes atrophy by disuse and osteoporosis,⁴ as well as preventing the rapid progression of scoliosis⁵ and allowing the patient more independence.⁶

In an attempt to prolong ambulation, a well-known resource prescribed to patients with DMD is the use of orthoses for the lower limbs.^{7,8} The main types mentioned for this purpose are the *Knee-ankle-foot* orthoses (KAFO),⁹ also called long¹⁰ and suropodalic orthoses by some authors or *Ankle-foot* orthoses (AFO).¹¹ According to the "Guidelines for exercise and orthoses in children with neuromuscular disorders,"⁷ ambulating patients are recommended to use the KAFO during the day and the AFO during the night; patients in wheelchairs are recommended to use the AFO only during the night. Despite these directives, some questions remain unanswered such as (1) the ideal moment to prescribe, (2) the number of hours to use the device during the day and night, and (3) the medium and long-term effects of using these devices.

Bakker et al.¹ carried out a specific review on the effect of the KAFO and affirmed that the diversity of methods used by the researchers made it difficult to reach reliable conclusions on the use of this orthosis.¹ Recently, a Brazilian study reviewed the importance of orthoses for the lower limbs on DMD sufferers and concluded that the KAFO is the most used orthosis because it minimizes muscle contracture and deformities, prolonging ambulation.¹² Despite those findings, Fernandes et al.¹² included no studies that associated the use of orthoses with non-surgical interventions and, in some studies there is no identification of the type of orthosis used. Still, the prescription criteria for the use of the device were not explored nor were the evaluation criteria for its effect.

In addition, there is the fact that there are no reviews specifically evaluating the effect that using orthoses may have on the age at which the loss of mobility occurs. Thus, in view of the benefits of prolonging ambulation for these patients and of the heterogeneity of methods and results pointed out in different studies.

OBJECTIVE

The present work sought to make a literature review to verify the effect of using orthoses vis-à-vis prolonging ambulation in patients with DMD. Another goal of this study was to identify the prescription criteria for the use of the device, associated therapeutic interventions, and evaluation criteria for the effect of using orthoses.

METHOD

This review article was prepared in three phases: (1) pre-selection of potential articles, (2) selection of articles, and reading, and (3) synthesis of the findings. In the pre-selection phase, a bibliographical survey was conducted on the PUBMED, PEDRO, and SCIELO databases by using the keywords: *Orthoses, bracing, gait, gait loss, ambulation, Duchenne muscular Dystrophy*, and pre-selecting the abstracts of the articles whose titles were related to the search theme.

In the selection phase, the abstracts were read and those that were suited for the search theme were selected. The articles selected were analyzed investigating the following information: the type of orthosis prescribed, the time of the orthosis prescription, period of use (day or night), and the effect of using the device. In the third phase, the information that follows was compiled.

This review included original articles and case reports that approached their interventions with some type of orthosis, associated or not with other treatment strategies. This review excluded review articles, articles that discussed intervention with orthoses in diseases other than DMD, and articles that intervened in DMD without using orthoses or that did not report the type of orthosis used.

RESULTS

In the pre-selection phase, 34 abstracts were identified, but only 21 scientific articles were selected. Two manuscripts could not be accessed as a whole, and another five articles were dismissed because they presented no relevant data. Therefore, in the present study, 14 articles have been analyzed in their entirety and whose main information is presented in Chart 1.

The publication of the included articles ranged from 1962 to 2011. In relation to the samples they examined, the number of par-

ticipants varied between 1 and 144 patients, and the ages of the subjects varied between 4 and 31 years. The characterization of the sample articles was heterogeneous: some authors included information on the stage of the disease, others on the gait capacity, and others on the presence and intensity of muscle contracture or muscle strength.

In relation to their intervention with orthoses, all the studies used these devices (regardless of the type) associated with other therapeutic interventions such as orthopedic surgery, passive muscle stretch, use of corticoids, physical therapy treatment, and physical exercise. Considering the type of orthosis and the period of use, 11 of the 14 selected articles conducted interventions with the use of the KAFO during the day; two articles evaluated the effect of using the AFO at night, and one article evaluated the effect of the AFO, considering both those patients who used it during the night and those who used it during the day.

In relation to the effects of using an orthosis, only one study failed to report the effect of using the device on ambulation. All the remaining articles had results showing that the orthosis delays the appearance of muscle contracture and prolongs ambulation.

The criteria for the prescription of the orthosis, as well as the variables evaluated to verify its effect, regardless of the type, were quite diverse. The frequency of falls, age, and mobility of the patients were some of the criteria used for the prescription. Among the variables evaluated to verify the effect of this device, muscle strength, the presence of muscle contracture, and functional capacity were all noteworthy.

DISCUSSION

The prescription of orthoses for the lower limbs of patients with DMD and the result of such an intervention are influenced by various factors such as the choice of type of orthosis, clinical characteristics of the patient at the moment of evaluation and prescription, and characteristics of the intervention the patient has been receiving.⁶ In view of this variety of factors, many studies sought to identify the effect of using this device, however, the methodological variety present in those studies makes it difficult to compare them.

It is known that the nocturnal and continuous use of the AFO, when initiated early, can delay the loss of the mobility by delaying the development of muscle contracture.¹¹ In

Chart 1. Types and results of orthotic interventions in patients with Duchenne Muscle Dystrophy

	Type of orthosis	Criteria for the prescription of the orthosis	Period the orthosis was used	Treatment associated with the orthosis	Variables evaluated	Effect of using the orthosis	Number of patients (age)
Williams et al. ¹⁰ (1984)	Long orthosis and AFO	Type of orthosis according to the patient's mobility	Nocturnal AFO: independent gait Diurnal long orthosis and nocturnal AFO: pre-loss of gait Diurnal AFO: people in wheelchairs	Surgery Physiotherapy	General mobility Muscle power Muscle contracture Pulmonary function Scoliosis	It prolongs ambulation due to the stretching of calf muscles	69 (4 to 17 years)
Spencer, Vignos, ⁴ (1962)	Long orthosis	Cannot walk independently	Diurnal (at least 3 hours/day)	Surgery Physiotherapy	Muscle strength	It allows ambulation even with muscle weakness	15 (8 to 13 years)
Hyde et al. ¹³ (2000)	AFO	Age (> 4 years) and walking independently	Nocturnal	Stretching	Anthropometric measurements Muscle strength Muscle contracture	It promotes stretching It delays the appearance of muscle contracture	27 (> 4 years)
Pardo et al. ¹⁴ (2011)	Long orthosis	16 years	Diurnal	Corticoids Surgery	Clinical evaluation (not specified)	It prolongs ambulation associated with the use of corticoids	1 (20 years)
Vignos et al. ⁹ (1983)	KAFO	Cannot walk independently	Diurnal	Surgery Physiotherapy	Vital capacity Creatinine coefficient	Not reported	50 (average of 6.7 years)
Seeger, Caudrey Little ¹⁵ (1985)	AFO	Age (> 5 years)	Nocturnal/diurnal	Surgery Stretching	Passive range of joint movement	It promotes stretching It reduces the equinus deformity	1 (5 to 16 years)
Vignos et al. ¹⁶ (1996)	KAFO	Contracture of the calcaneal tendon (1.5 + - 2.8 degrees, range of 0 to 8 degrees)	Diurnal	Passive stretching Surgery	Weight and height Vital capacity Creatinine indices Muscle strength	It prolongs ambulation by preventing muscle contracture	144 (6 to 31 years)
Heckmatt et al. ⁶ (1985)	KAFO	Cannot walk independently	Diurnal	Surgery Physiotherapy	Physical performance and social factors (not specified)	It prolongs ambulation and delays appearance of scoliosis	57 (6 to 13 years)
Rodillo et al. ⁵ (1988)	KAFO	Between 6 and 12 years	Diurnal	Physical exercise (walking)	Scoliosis	It impedes the rapid progression of scoliosis	93 (6 to 12 years)
Scott et al. ¹¹ (1981)	AFO	Between 4 and 12 years	Nocturnal	Stretching	Functional capacity Muscle contracture	It delays the appearance of muscle contracture of the calcaneal tendon and thus, delays the loss of ankle dorsiflexion	59 (4 to 12 years)
Hyde et al. ¹⁷ (1982)	KAFO	Cannot walk independently and suffers many falls	Diurnal	Surgery Physiotherapy	Functional capacity Muscle strength Physical performance Passive range of joint movement	It prolongs ambulation	30 (7 to 13 years)
Melkonian et al. ¹⁸ (1980)	KAFO	Increase in the frequency of falls	Diurnal	Surgery	Muscular activity	It prolongs ambulation, for the muscle activity becomes minimal with	13 (average of 10 years)
Taktak, Bowker, ¹⁹ (1995)	KAFO	Age (> 9 years)	Diurnal	Physical exercise	Weight of the orthoses Velocity and energy expenditure during gait	The new orthosis model promoted less energy expenditure and increase in speed	18 (7 to 12 years)

KAFO: Knee-Ankle-Foot Orthosis; AFO: Ankle-Foot Orthosis.

this review, the effect of using the AFO was evaluated in only three of the 14 articles included, with two of these articles having the prescription of nocturnal use for all the patients^{11,17} and in one article, patients who used it at night and patients who used it during the day were evaluated together.¹⁵

According to the study by Seeger et al.¹⁵ the rate of progression of the equinus deformity is, on average, 0.38° per month, with the slowest progression having been associated with the greater frequency of nocturnal use of the AFO and the more rapid progression was associated with the interruption of noc-

turnal use of the device. The evaluation of the effect of the AFO seems to occur always under the view of preventing muscle contracture and ankle deformities. However, the effect of the AFO during ambulation or from the biomechanical point of view has not been investigated yet.

According to the results of the present review, the KAFO was the type of orthosis most used, with this device being effective at prolonging ambulation in patients with DMD, when prescribed in the phase that precedes the loss of gait. It is noteworthy that, in 6 of the 11 studies that discussed the use of the KAFO, a surgical intervention was made to correct deformities before beginning the use of the device. After surgical correction, the use of the KAFO promoted better alignment of lower limbs, minimizing the advance of muscle contracture in flexion.⁴ Despite these benefits, the additional weight of the long orthoses may be an important factor in explaining the fact that this type of device is not popular among Brazilian patients.²⁰

Although there are no kinematic and kinetic studies about the use of the KAFO, a few biomechanical considerations about the effect this type of orthosis has on ambulation have already been made. The use of the KAFO favors the initial contact with the heel, but impedes the flexion of the knee, compromising the balance phase in the gait. The initial contact made with the heel allows the anterior progression of the limb. However, the KAFO maintains an anterior support on the tibia that impedes the flexion of the knee. Thus, the patient continues to use the strategy of lateral inclination of the trunk so that the lower limb can advance. The use of the KAFO allows for a safe ambulation by guaranteeing the stability of the lower limbs, however, even with the use of the device, the hip joint stability is still maintained due to the lordotic posture and to the posterior inclination of the trunk.²¹

For patients with DMD, prescriptions for the KAFO have been favored over the AFO, because the AFO has an indirect action on the knee, while the KAFO maintains the alignment of this joint directly.²¹ According to these authors, the muscle strength of the knee and hip in patients with DMD is inadequate to control the action of the knee joint during gait, even with the support of an AFO. Thus, as detailed before, the KAFO provides an anterior support on the tibia, preventing the flexion of the knee and favoring an appropriate position of the foot. In addition to these effects, the KAFO reduces the electrical activity of the lower limb muscles during gait, which may indicate that this device eliminates the need for activating the muscles evaluated.¹⁸

Despite these biomechanical suppositions on the use of the KAFO, the ideal moment to prescribe it is still controversial. In the studies by Hyde et al.¹⁷ and by Vignos et al.,⁹ the KAFO was prescribed when the patients started to

fall more often and could not walk long distances independently. In parallel, Heckmatt et al.⁶ prescribed the KAFO when the patients had already lost their capacity to walk functional distances, but still were able to maintain orthostatism and to walk a few steps. In view of that, it can be observed that the prescription criteria for the device are usually subjective. The most objective prescription found in the literature was the one in a study by Vignos et al.¹⁶ in which the exclusive intervention with the KAFO was prescribed when the patients showed a slight muscle contracture of the calcaneal tendon (average and standard deviation 1.5° and 2.8°, respectively). However, the patients who underwent surgical intervention before using orthosis showed a dorsiflexion range of motion of 10° after surgery (moment of intervention with the KAFO).¹⁶

The results of the present review indicated that the period of use of the devices followed the directives of the "Guidelines for exercise and orthoses in children with neuromuscular disorders," in which orthoses like the KAFO can be prescribed for diurnal use and like AFO for nocturnal use.⁷ Despite these directives, the daily hour load of using the device is an aspect rarely discussed in studies. Of the articles inserted here, only one detailed this variable, prescribing a minimum time of 3 hours/day for the use of the KAFO, and concluded that it allows for ambulation in spite of muscle weakness.⁴

In general, the use of orthoses, regardless of the type, can delay the age at which one loses ambulation by delaying the advancement of muscle contracture. In addition, orthoses have always been used in association with other types of therapeutic interventions. Hyde et al.¹³ for example, reinforced that to control the muscle contracture in plantar flexion it is more beneficial to associate the orthosis with daily stretching as a therapy than only doing the stretching. Still, according to Seeger et al.,¹⁵ the progression of the equinus deformity is more rapid in patients who did not receive surgical intervention. Thus, the need has been verified to associate the use of orthosis with another therapeutic intervention such as the use of corticoids,¹⁴ surgical correction,^{4,10,18} passive stretching,^{11,17} or physical therapy treatment.^{4,10}

Chart 1 shows that the evaluation of the effect of using orthoses can be made through various variables-the most relevant ones in clinical practice being range of motion, muscle strength, and motor ability. Although many studies evaluated the effect of using orthoses, important questions still remain to be clarified

concerning the prescription and biomechanical and energetic effects of these devices on the gait of patients with DMD.

CONCLUSION

The use of orthoses, regardless of the type, seems to delay the age at which a patient with DMD loses ambulation, for it minimizes the advancement of muscle contracture, even in the absence of surgical intervention. The KAFO is commonly prescribed for the ambulation phase that precedes the loss of gait, whether or not associated with the surgical correction of deformities. The surveyed data recommend the late use of the KAFO for ambulation and the early nocturnal use of the AFO to minimize the progression of deformities and, consequently, allow for the possibility of a future intervention by the KAFO not associated with any surgical intervention. In parallel, future scientific studies should evaluate the biomechanical effects and those of energetic expenditures of using the AFO and KAFO during ambulation in this population.

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