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Serial Casting for the Management of Ankle Contracture in Duchenne Muscular Dystrophy

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Abstract

Purpose—To evaluate the effect of serial casting in boys with Duchenne muscular dystrophy.

Methods—Chart review of 9 patients with Duchenne muscular dystrophy, mean age 8.9 ($\pm$ 2.1) years.

Results—Initial dorsiflexion – 6.2° and – 5.2° right and left, respectively. The mean improvement was 12° and 11.6° on the right and left (knee extended) and 7.7° and 8.7° on the right and left (knee flexed) or 2.7° and 3.9° per cast, respectively. Times to run 10 m, climb 4 steps, and get off the floor were unchanged. Correlations between range-of-motion change/cast and age were $r = -0.86$ right and $r = -0.84$ left. Three patients had delayed onset foot pain; one child had redness with symptom resolution in all cases.

Conclusions—Improvement in range of motion with the application of serial casting was found with no loss of function or speed despite the period of immobilization.

Keywords

ankle; child; contracture; Duchenne muscular dystrophy; fiberglass casts; motor activity; passive range of motion; physical therapy/methods

INTRODUCTION

Duchenne muscular dystrophy (DMD) is one of the most common childhood muscle disorders occurring in 1 in 3500 live male births,¹ and results from a mutation of the dystrophin gene.² This genetic mutation results in the absence of the dystrophin protein,³ an integral component of the functional connection between the actin and myosin contractile apparatus in the muscle cell and the sarcolemma of the muscle cell. This form of muscular dystrophy occurs almost exclusively in boys because of the location of the dystrophin gene on the X chromosome. Boys with DMD demonstrate motor development that is marked by

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an initial period of gain in motor skill, followed by a plateau in skill acquisition that is then followed by a decline in motor skills and an ultimate loss of ambulation. Corticosteroid treatment has been shown to modestly prolong ambulation and is the only therapy available for the progressive weakness underlying the loss of ambulation these children experience.⁴ Impairments of body structure and function include the development of weakness and the formation of contractures that initially are apparent in the gastrocnemius complex and later develop in the hamstrings, hip flexors, and tensor fasciae latae musculature. The development of contractures, together with the progressive weakness, creates a standing posture that is marked by plantar flexion, anterior pelvic tilt, and hyperlordosis. This posture allows the weight line to fall anterior to the knee and posterior to the hip, enhancing the typical stance stability of the body. In addition, knee stability is aided biomechanically by the ground reaction force of the gastrocnemius. This force is transmitted from the contact point with the ground by limited dorsiflexion creating an extension moment at the knee by restraining the forward progression of the tibia.⁵ The torque at the ankle that can contribute to this knee moment is related to the distance that the force acts (the contact point with the floor) from the axis of the ankle. As the gastrocnemius becomes contracted and the child walks progressively more and more on his toes, this distance becomes shorter and shorter and contributes less and less force to the knee extension moment.⁶ In addition, once the heel no longer contacts the floor, the base of support becomes shortened and the child's cone of stability is diminished, making maintenance of standing balance increasingly difficult.

In the past, maintenance of muscle length has been addressed by orthopedic surgery. This treatment has generally been applied mainly in 1 of 2 ways. In the first, surgery is done at the end stages of ambulation, to prolong walking through the use of knee-ankle-foot orthoses (KAFOs). In the second approach, surgery is done earlier in the course of the disease with the idea of prolonging brace free ambulation.⁷ Overlengthening and ambulation loss with this earlier approach to surgical lengthening of the gastrocnemius complex have been reported. In addition, surgical lengthening is often followed by the use of KAFOs to maintain ambulation. These braces often limit the ability of a boy with DMD to attain standing independently because of the weight of the brace and the necessity of having the knee locked. While some centers favor surgical lengthening at the end stages of ambulation to prolong the ability to walk, this has not been uniformly recommended.⁸

Conservative treatments aimed at preventing contracture formation and equinovarus deformities of the ankle include stretching and nighttime splinting with ankle foot orthotics. Use of night splints, in combination with daily stretching of the heel cords, plays a significant role in preventing the development of an equinovarus deformity at the ankle.⁹ The use of night splints has been shown to be more effective at preventing contracture of the Achilles tendon than stretching alone.¹⁰ Boys with DMD who have used night splints prior to the loss of ambulation walked independently longer than those who did not use splints.¹¹ Despite the benefits of using these conservative methods, contractures continue to develop in boys with DMD as the disease progresses. When this occurs, night splints are often not tolerated well. Stretching and wearing night splints work well to prevent contractures; however, once a plantar flexion contracture is present. These methods typically are not used to regain motion at the joint; rather, they are used primarily to decrease the rate of contracture progression.

Serial casting is one method that has been used to increase the length of muscle in other populations of children who have a tendency to form contractures, most notably in children with cerebral palsy (CP). In animal models, muscles respond to immobilization in a lengthened position with both an increase in the number of sarcomeres and by stretch of the tendon. These 2 processes are age dependent. In immature muscle, the ratio of tendon stretch to sarcomere growth is increased when casting is undertaken. At the onset of immobilization in animal models, immature muscle responds with sarcomere growth but after about 5 days of immobilization, tendon stretch progressively replaces sarcomere growth. Adult muscle in animal models responds to longer immobilization primarily with sarcomere growth and with only a limited amount of tendon stretch.^{12–16} In the mdx mouse, an animal model with similar symptomatology to DMD, the increase in the number of sarcomeres noted after limb immobilization in a lengthened position is delayed.

A significant increase in the number of sarcomeres in the mdx mouse is noted only after 10 days of immobilization in a lengthened position as compared to 4 days for normal muscle.¹⁷ A significant literature base supports the use of serial casting in children with CP to improve muscle length and to decrease spasticity. Serial casting has been combined with or compared to botulinum toxin in a number of studies and serial casting has been shown to improve ankle kinematics during gait, spasticity, passive range of motion (ROM), and dorsiflexion strength as compared to baseline in the population with CP. However, most of the literature focuses on the addition of botulinum toxin to the casting treatment and not directly on the effect of casting alone versus no treatment.^{18–25}

In individuals with neuromuscular diseases, the literature is significantly more limited. Rose et al²⁶ evaluated the effect of night casting in a sample of individuals with Charcot Marie Tooth disease and demonstrated a 4° improvement in ROM as compared with a control group. Main et al²⁷ and the group at Hammersmith Hospital have reported serial casting boys with DMD. Their goal was to initiate KAFO use toward the end of independent ambulation. With the use of glucocorticoids, boys with DMD continue to walk longer but the prevention of contractures does not appear to result from this therapy to the degree that muscle strength improves.²⁸

Despite the successful use of serial casting in populations such as children with CP, the use of serial casting in boys with DMD is not widely accepted. This may be due to the lack of supporting literature. The goal of serial casting in this study was to improve passive dorsiflexion and prevent a decline in motor function. We have casted children earlier in the course of the disease, as compared with Main and colleagues,²⁷ with the goal of maintaining brace-free ambulation. Our policy has been to cast children when they no longer demonstrate dorsiflexion to neutral with the knee extended and when they were still able to rise from the floor independently, using a Gower maneuver. We limited our application of casting to those boys with relatively mild limitations of activity who were still able to rise from the floor because of the more tenuous status of ambulation in those who are not able to complete this task. The purpose of our retrospective study was to describe our initial experience using serial casting in boys with DMD in the setting of a tertiary care specialty clinic, to quantify our results, assess the risks, and demonstrate the benefits of casting this population.

METHODS

The study design was a case series with retrospective chart review that followed institutional review board approval. Inclusion criteria included boys with DMD confirmed by biopsy or dystrophin gene mutation who underwent serial casting within the study window. Boys were generally considered appropriate for casting if they were able to rise from the floor independently using the Gower maneuver and lacked ROM to neutral dorsiflexion and/or were ambulating habitually on their toes. The records of 9 boys with DMD with a mean age of 8.9 years (± 2.1) were included. Initial dorsiflexion ROM at the ankle was -6.2° (± 5.7) on the right and -5.2° (± 3.9) on the left measured in the supine position with the knee extended and -0.2° (± 6.3) on the right and -1.4° (± 5.2) on the left measured in the supine position with the knee flexed.

Serial casts were applied by a physical therapist (A.G., J.F., or K.D.) trained in a standard method, with the child placed in the prone position with the knee in 90° of flexion. The foot and lower leg were covered with stockinet and bony prominences were padded with under cast padding. Fiberglass casts were applied with the ankle at a comfortable end range of dorsiflexion and with the midfoot in neutral alignment. Posting of the heel was used as needed to ensure that the child was able to walk with a plantigrade gait without an assistive device. Casts were changed once each week with the new cast placed in greater dorsiflexion depending on ROM available. Casts were reapplied immediately following timed testing and ROM measurement. The period of casting was determined by the clinical needs of each child and ranged from 2 to 5 casts with a mean of 3 casts. During the period of casting, we ensured that each child was able to continue to ambulate with cast boots at home and in the community.

Serial casting outcomes were monitored by measures of activity including timed tests that were performed before the onset of casting and after each serial cast was removed. These timed tests included the time to rise from the supine position to the standing position (Gower maneuver), 10-m walk/run, and time up 4 steps. These methods were completed in a standardized manner with a stopwatch and the same equipment; 4 wood steps with rail, 10-m course with marked distance, and a firm mat where the child was placed in a supine position and asked to "get up as quickly as you can." Measurements of impairment of body structure and function included passive ROM in dorsiflexion by standard goniometric technique²⁹ with the knee in a flexed ($n = 6$) and extended ($n = 9$) position by a physical therapist with more than 10 years' experience (A.G., J.F., or K.D.).

RESULTS

The mean change in ankle dorsiflexion ROM with the knee extended was 12° (± 5.5) and 11.6° (± 6.2) on the right and left, respectively. Notable was the improvement from -6.2° and -5.2° of dorsiflexion to $+5.8^\circ$ and $+6.3^\circ$ of dorsiflexion on the right and left, respectively. With the knee flexed, dorsiflexion improvement was 7.7° (± 7.2) and 8.7° (± 5.8) on the right and left, respectively (Table 1). The number of casts per boy varied with a mean per cast improvement of 2.7° (knee flexed) and 3.9° in ROM over the average 3 cast course of treatment. No significant change in timed motor function tests was noted in the

time taken to walk 10 m, climb 4 steps, and get off the floor with pre- and postcasting times highly correlated (Table 2). There was a significant negative correlation between ROM gain per cast and age ($r = -0.86$ on the right and $r = -0.84$ on the left). The younger boys demonstrated a more rapid improvement in ROM than the older boys. Of the 9 boys (18 ankles) who were cast, 3 had delayed onset foot pain (either midfoot or Achilles tendon) with onset 3 days after casting. All symptoms resolved either spontaneously or with the use of nonsteroidal anti-inflammatory medication and no recurrence was noted. One child had focal redness of the skin within the cast area, which resolved spontaneously but delayed application of the next cast for 1 week.

DISCUSSION

Similar to children with other diseases, boys with DMD demonstrate improvement in ROM at the ankle with the application of serial casting. Our results suggest that measures of activity remain stable following serial casting with no loss of function or speed noted despite the period of immobilization. The loss of ambulation in boys with DMD primarily arises as a result of a combination of factors including progressive muscle weakness, contractures, and likely the increased body size that occurs with growth. Steroid therapy has been commonly applied in this population and is effective in addressing muscle weakness. However, our options for improving contracture are limited and until now have only been effective at slowing the progression of contractures. This and the work of Main et al²⁷ are the first nonsurgical attempts to address contractures by serial casting with the goal of improving flexibility and maintaining ambulation and functional skills.

Stability of timed motor function testing over the course of casting is an important factor in this population, given that the treatment presented the potential for atrophy of the immobilized muscles. The goal of the treatment was directed at ROM and we did not expect to see an improvement in functional skills. A prospective study of longer duration employing a control group with no treatment could reveal the effect of ROM improvement on activity limitations, as well as the degree to which ROM gains could be maintained over time. Timed testing does not capture a change in the quality of gait mechanics; however, other improvements in gait could have been experienced by the boys such as improved heel contact during the stance phase of gait although this was not formally measured. Additional outcome measures including instrumented gait analysis and energy expenditure would be of interest in further examining these parameters.

Even though the children were not asked to wear knee immobilizers at night to selectively stretch the gastrocnemius, results point to a greater increase attributed to the gastrocnemius as compared to the soleus. This may have resulted from the additional stretch that occurred with knee extension while walking and the fact that the ankle was cast with the knee in flexion, which placed the gastrocnemius in a slack position at the knee during cast application. The addition of knee immobilizers at night with serial casting may provide additional improvement in ankle ROM and may warrant additional study.

Data from various animal models show that the gain in ROM results from a combination of tendon stretch and increase in the number of sarcomeres in series within the muscle. In older

animals, there is a higher proportion of intramuscular sarcomere addition as compared to tendon stretch.^{14,15,17} Our sample demonstrated a negative correlation between ROM gain per cast and the child's age. A per cast outcome was used because in the context of this retrospective study the number of casts applied varied from boy to boy and was dependent on the degree of ROM correction that was needed. This correlation between ROM improvement and age could reflect the effect of increased levels of intramuscular fat and fibrosis resulting in decreased sarcomere adaptation or decreased elasticity of the tendon in the older boys with DMD. Despite this, our population demonstrated improvements in dorsiflexion with both the knee flexed and extended reflecting an increase in the overall length of the musculotendinous unit.

We have demonstrated in our sample of boys with DMD that improvement in ROM was possible with the application of serial casting and that activity, as represented by our timed testing outcome, remained stable during the period of casting. A longer-term controlled study is needed to determine whether serial casting provides long-term advantage for impairments of body structure and function, including hypoextensible muscles, and advantage for activities such as walking and stair climbing. In addition, the characteristics of the casting program in terms of length of cast application, criteria for initiation of a casting program, and optimal characteristics of the casting process need to be determined.

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TABLE 1

Range of Motion

	Right Dorsiflexion Knee Extended (n = 9)	Left Dorsiflexion Knee Extended (n = 9)	Right Dorsiflexion Knee Flexed (n = 6)	Left Dorsiflexion Knee Flexed (n = 6)
Mean initial ROM (SD)	– 6.2 (5.7)	– 5.2 (3.9)	– 0.17 (6.3)	0 (4.0)
Mean final ROM (SD)	5.8 (4.8)	6.3 (4.5)	7.5 (6.7)	8.7 (5.2)
Mean change (SD)	12.0 (5.5)	11.60 (6.2)	7.7 (7.2)	8.70 (5.8)
<i>P</i> (95% CI)	<0.0001 (7.8–16.2)	0.001 (6.8–16.8)	0.047 (– 0.1 to 15.2)	0.014 (2.6–14.7)

Abbreviations: CI, confidence interval; ROM, range of motion.

TABLE 2

Timed Testing

	10-m Run Time	Timed Gower	Time Up 4 Steps
Initial time mean (SD)	5.7 (1.4)	5.4 (2.4)	4.2 (1.5)
Final time mean (SD)	5.9 (1.4)	5.1 (1.8)	4.3 (1.7)
Change mean (SD)	0.2 (0.5)	−0.31 (1.0)	0.05 (0.5)
Correlation	0.930 ($P=0.0001$)	0.921 ($P=0.0001$)	0.954 ($P=0.0001$)
$P(95\% \text{ CI})$	0.254 (− 0.2 to 0.6)	0.394 (− 0.5 to 1.1)	0.813 (− 0.4 to 4.9)

Abbreviation: CI, confidence interval.