

Lower Extremity Review

# ler:PEDIATRICS

February 2018



**Update: Orthotic care and physical therapy for DMD**

**Media, toys, and games for kids with disabilities**

**PLUS:**

- Pitching and lower extremity recovery
- Autism linked to motor impairments
- Ankle strength and gait profiles in CMT

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### 15 Media, toys, and games for kids with disabilities

Children use toys and media characters to spark their imaginations and cast themselves as the star of their own stories. Through play, they connect with other kids and dream about their future. Options are few, however, for kids who wear a device or otherwise look "different" to see themselves reflected during play.

By Jill R. Dorson

## From the editor: Reflections on self-image



The first article I wrote for any *LER* publication examined the negative impact a therapeutic device can have on self-image, and how this can create a major barrier to patient satisfaction and adherence. (See "Sensitivity to self-image boosts O&P outcomes," *LER*, April 2011.)

The article, which focused on adults, looked at findings of research and practitioners' anecdotal experience that revealed how patients' feelings about their device—more accurately, their feelings about themselves when they wear a visible orthosis or prosthesis—govern how likely they are to use the device as prescribed.

In children, who are still developing self-identity, viewing themselves, or being viewed by others, as "different" likely has even more of an unhelpful impact on self-image—and on adherence—than it does in adults. Why? One reason is the relative scarcity of media characters and toys with which kids who have a visible lower extremity condition can identify.

In "Media, toys, and games for kids with disabilities," on page 15, writer Jill Dorson details one child's campaign to see more kids like her reflected in toys, on bookshelves, and in other media, as well as three orthotic technicians' contribution to the lack of such depictions: a storybook called "Beau and His New AFO."

Their book is a great resource. More like it are needed, but if you don't have time to write and illustrate one, there are still ways to help.

Children are likely to feel better about a device when they have a voice in how it looks, according to a survey of pediatric patients at Shriners Hospital for Children—Tampa in Florida. Results showed that children who had an orthosis with patterns and colors they'd selected had a more positive self-image, better peer acceptance, and, consequently, improved adherence, compared with peers who wore white braces.

In the digital age, orthotic and prosthetic personalization can be done quickly. The psychological payoff for kids—feeling better about themselves and their device—can be long-lasting.

Emily Delzell, *Editor*

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# Pitchers don't get enough time for lower extremity strength recovery

## Single-sport players need more rest days

By Katie Bell

Current Little League pitching guidelines provide insufficient lower extremity strength recovery time in youth pitchers, even when they are pitching at submaximal pitch counts, according to research from California published in January.

Study coauthor Jennifer Livingston, PhD, ATC, associate professor in the Department of Kinesiology at Azusa Pacific University in California, said, "In our pilot study, we didn't collect any data related to injuries, but I'd suspect fatigued legs would require the pitcher to rely more heavily on the upper extremity to generate momentum, placing the arms at greater risk of injury."

The study included 15 healthy male baseball pitchers (average age, 9 years) without a current injury or history of injury two months prior to the start of the study. Livingston and colleagues calculated the change in lower extremity force production after a pitching performance and over subsequent days of rest required for the pitchers to recover.

Using a handheld dynamometer, the researchers measured force production on the pivot leg and the stride leg for the gluteus maximus, hamstrings, gastrocnemius, gluteus medius, and quadriceps prior to pitching, immediately after pitching a submaximal number of pitches, and for the following four days. They collected the pitch velocity (PV) for each pitch and the rating of perceived exertion (RPE) immediately after each component to measure fatigue during performance.

They found that, on average, 47.8 pitches per set were thrown; the average PV was 20.4 m/s and the average RPE was 6.9. Twelve participants threw the assigned number of pitches before reaching 9 on the Pictorial Children's Effort Rating Table (PCERT), which provides subjective information of the participant's overall efforts and fatigue rate (based on scoring using a 1-10 scale, with "1" being "very very easy" and "10" being "so tired I'm going to stop." One child completed all assigned pitches and reported a PCERT of 9 at the session's end.

The researchers noted that, with the exception of maximum RPE, several factors were different between the four children aged 8 to 9 years and the 11 children aged 10 to 11 years. Older pitchers were heavier, taller, and had a higher fat-free mass than younger counterparts. The velocity and number of required rest days were higher in the older participants; their baseline strength was higher than strength

None of the boys had fully recovered by the time Little League guidelines would permit them to pitch again.

immediately after pitching in the stride leg for the hamstrings, triceps surae, gluteus medius, and quadriceps.

Older participants had a lower percentage of baseline strength than younger participants in all muscle groups during the post-test, suggesting older players rely more on the lower extremity during pitching, according to the study authors.

The researchers also compared the number of days required for each muscle group to return to baseline force production levels with Little League rest guidelines for pitchers.

"The reduction in force production and time to recover varied among participants and muscles, but none of the boys fully recovered by the time Little League guidelines would permit them to pitch again," said Livingston. "The number of muscles that failed to return to baseline strength ranged from one to all ten of the tested lower extremity muscles."

Livingston said the data support the idea that not all pitchers should be expected to throw to their limit set by the Little League, and that "the coaches, parents, and youth athletes should watch for signs of fatigue and build endurance first."



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Furthermore, the study found that the difference between mandated and actual required rest time was lower for participants who played other sports than for those who played only baseball.

Coauthor Nicholas Tavoukjian, MS, ATC, athletic trainer at Evergreen Physical Therapy Specialists, in Pasadena, CA, said, "The offseason should be taken advantage of; either by having kids play different sports that bias other muscle groups, or by including offseason strength training and conditioning. I think the model of playing Little League and travel ball concurrently is flawed and places kids at risk for overuse injury." (See "Early athletic specialization: Misconceptions and hazards," February 2017, page 15.)

Tavoukjian added, "I do not necessarily think the Little League guidelines should change. I do think there should be a greater emphasis on skilled strength training and conditioning for youth athletes to facilitate improved recovery and well-rounded athleticism."

Commenting on the study, Gretchen Oliver, PhD, ATC, CES, assistant professor of kinesiology at Auburn University in Alabama, noted that, because the study did not collect the pitchers' RPE on the final allotted day of rest, it's unclear whether pitchers were fatigued in their lower extremity at that time—but they did have decreased strength.

"There is no need to push the kids to fatigue," she said. "Get to know your pitchers, get to know their postures. Posture on the mound can say a lot." More research examining fatigue, measures of fatigue, and consecutive days of fatigue and performance is needed, Oliver added. 

*Katie Bell is a freelance writer based in New York City.*

Source:

Livingston JL, Tavoukjian NM. Lower extremity strength and recovery time in youth baseball pitchers: a pilot study. *J Strength Cond Res* 2018 Jan 29. [Epub ahead of print]

# Autism linked to between-limb asymmetries across the gait cycle

## Gait pattern ID could help target therapy

By Keith Loria

Recent findings from the University of Nevada, Las Vegas (UNLV), add to a growing body of evidence that autism spectrum disorder (ASD) is associated with significant gait asymmetry, suggesting movement quality should be part of the diagnostic and treatment processes for ASD.

In 10 children with ASD (4 girls) aged between 5 and 12 years, UNLV researchers found kinematic and kinetic between-limb asymmetries across the gait cycle. These included asymmetries in sagittal plane hip, knee, and ankle joint positions, as well as vertical and anterior-posterior ground reaction forces (GRF).

The findings, published last June in *Gait and Posture*, are consistent with those of other studies that have identified asymmetries in children with ASD at discrete time points during gait, said Jeffrey D. Eggleston, MS, first author of the study and a doctoral candidate in the Department of Kinesiology and Nutrition Sciences at UNLV.

"While there is still little known regarding gait impairments in children with ASD, our findings illustrate that gait descriptors may provide insight into furthering working knowledge of ASD and may even enable gait-related symptoms to be treatable through therapies and interventions," he said.

Despite growing research suggesting children with ASD exhibit some form of motor impairment, it is not used in the diagnostic criteria for ASD established by the American Psychiatric Association, and Eggleston said many children with ASD do not receive physical therapy interventions.

The types of asymmetries observed in the study could lead to biomechanical compensations with long-term clinical implications, he said.

"By way of example, individuals with hip joint arthritis or other anomalies, facing hip arthroplasty, often accommodate in the short term by limping during locomotion," Eggleston said. "While such compensatory strategies are useful in the short term, chronically they could lead to low back misalignment, strength imbalances, and/or balance and general locomotor challenges."

The Model Statistic gait analysis technique used by the researchers allowed

them to analyze asymmetries across the gait cycle and to identify subphases of gait when asymmetries are more prominent, according to Janet S. Dufek, PhD, professor and associate dean of the School of Allied Health Sciences at UNLV, and senior author of the study.

The UNLV researchers used the same analysis method as in a January 2017 study that confirmed significantly greater asymmetry in children with ASD than their typically developing peers.

In the current study, though, the extent to which asymmetries were associated with specific gait subphases varied between participants and didn't exhibit generalizable patterns in this small sample. The authors,

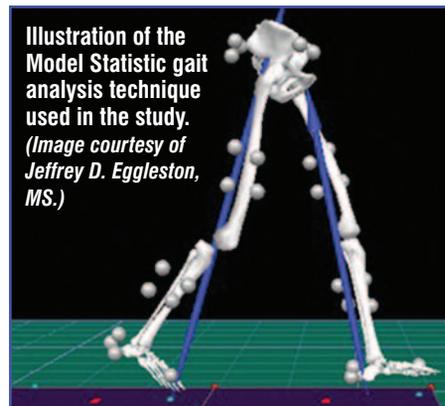
"Potential key contributors for gait asymmetry originate in the motor-controlling functions of the brain."  
—Janet Dufek, PhD

however, emphasized its potential utility for identifying specific gait issues in individual patients, which would facilitate more targeted therapy.

"Due to the highly heterogeneous nature of ASD and the variety of symptoms or comorbidities children and adults may have, it is paramount that any therapies be specific to the person and their own limitations," Dufek said. "While this is common in most rehabilitation settings, therapists seeing patients with ASD to address motor challenges may not be the norm. It is suggested that as such, an interdisciplinary approach to treatment may be the best approach."

Additionally, the percentages of observed GRF asymmetries were much lower than the percentages of kinematic asymmetries, which could also have clinical implications as it suggests that segmental control may be more important than propulsive and braking abilities in this population.

Dawn-Marie Ickes, MPT, PMA-CPT, an assistant professor in the Department of



Physical Therapy at Mount Saint Mary's University in Los Angeles, said the biggest influence she sees on gait symmetry and ASD is the integration of the individual's sensory processing with their proprioception.

"PT should be modified to accommodate the special needs of children with ASD in that it should take a multifaceted approach to the assessment of functional movement patterns as they relate to gait to determine how to best balance the asymmetries which present during the gait cycle," she said. "A combination of movement-based therapeutic interventions selected based on the client's unique presentation is a good place to start, and early modulation and integration of the vestibular system in the treatment session has worked for increased treatment tolerance in my practice."

The UNLV researchers noted the complex nature of ASD makes it difficult to explain the mechanisms underlying their findings, but one possibility is that children with ASD lack the coordinated movement patterns that allow for symmetrical gait.

"Alternative hypotheses suggest that children with ASD exhibit dysfunctional segregation of the motor cortex, which may be the key to uncoordinated movements," Dufek said. "Additionally, there is the potential for the cerebellum, which receives sensory information and regulates movements, to have a level of dysfunction as well. Viewed collectively, the potential key contributors for gait asymmetry originate in the brain and specifically, the motor-controlling functions of the brain." 

Keith Loria is a freelance writer in northern Virginia.

Sources:

Eggleston JD, Harry JR, Hickman R, Dufek JS. Analysis of gait symmetry during overground walking in children with autism spectrum disorder. *Gait Posture* 2017;55:162-166.

Dufek JS, Eggleston JD, Harry JR, Hickman R. A comparative evaluation of gait between children with autism and typically developing matched controls. *Med Sci* 2017;5:1.

# Measures of ankle weakness reveal varied CMT gait patterns

## Gait profile should guide orthotic design

By Keith Loria

Gait-pattern classification derived from functional measures of ankle weakness can be used to assess progression of Charcot-Marie-Tooth (CMT) disease and to guide clinical decision-making about treatment strategies, such as orthotic design and strength-training programs, according to a study from the University of Sydney in Australia, published in 2017.

Using 3D motion analysis, researchers examined gait patterns in 60 children with CMT aged 6 to 17 years (26 girls, 34 boys) and assessed differences between them and 50 healthy controls based on the CMT Pediatric Scale of functional ankle dorsiflexion and plantar flexion weakness.

Researchers split data into subgroups denoting increasing severity of dorsiflexion and plantar flexion weakness and found three distinct gait patterns at the ankle in kids with CMT: a near-normal gait pattern, with the only difference between kids with CMT and controls being a reduced maximum ankle dorsiflexion in stance; reduced maximum ankle dorsiflexion in swing (foot drop); and increased peak dorsiflexion and reduced ankle power generation. The third, most severely affected group, also had an increased mean sagittal plane knee extensor moment, indicating ground reaction force remaining behind the knee throughout stance.

Contrary to previous studies, there were no signs of reduced ankle power or compensation through “steppage gait” in this population with mild to moderate CMT.

“The paper shows many individuals with CMT do not fit a specific gait profile, and that’s significant to orthotics because it’s so critical to do proper evaluations of their gait and of their needs to develop a proper device plan,” David B. Misener, CPO, who practices with Clinical Prosthetics and Orthotics in Albany, NY, told *LER: Pediatrics*.

Long-term follow-up, for example, Misener said, is needed for children who fall into the study’s third group to monitor for development of crouch gait, which could occur because of the increasing ankle dorsiflexion, knee flexion, and knee extensor moment in stance.

All children with CMT had a significantly worse gait profile score than controls. Primary deviations at the ankle showed significantly reduced ankle dorsiflexion in

stance, reduced maximum ankle dorsiflexion during swing and at initial contact, reduced ankle dorsiflexion moment in loading response, and reduced external thigh-foot angle in midstance. There were no signs of reduced ankle plantar flexion or ankle power generation at push-off.

Compared with controls, children with CMT also demonstrated a significantly reduced external thigh-foot angle in the transverse plane, indicating altered foot alignment. Deviations at the knee included increased knee flexion at initial contact, increased peak knee flexion in loading response, and increased maximum knee flexor moment in single support compared with controls. At the hip, children with CMT showed reduced hip abductor moment in terminal stance and reduced internal hip rotation during midstance.

“Kids vary considerably in weakness in dorsiflexors early in CMT and in plantar flexors later on.”

—Ken Cornell, CO

Misener noted that the study recognized loss of range of motion (ROM), and said tight plantar flexion seems to be common yet sometimes overlooked in CMT, though the finding is critical from a therapy and patient awareness perspective. “Individuals can be trained by physical therapy to stretch tight muscles and tendons,” he said. “From a bracing perspective, these tight muscles and tendons can be accommodated within a custom insert or AFO.”

Ken Cornell, CO, who practices with Cornell Orthotics & Prosthetics in Peabody, MA, said orthotists need information like that provided by the study, which was published last July in *Gait & Posture*, to understand the needs of children with CMT.

“This study certainly proves we can no longer simply expect our CMT children aged eight to fourteen to have simple foot drop with progressing classic equinovarus foot and ankle deformities based on their diagnosis,” Cornell said. “They will vary con-



siderably in weakness in dorsiflexors early on in the disease and plantar flexors later on. Each patient will require a thorough evaluation, and classification should be based on dorsiflexion and plantar flexion function.”

Cornell points out that, since the 1980s, physicians have known that muscles innervated by the longest peripheral nerve axons are affected first in CMT patients.

“These types of conditions are now commonly referred to as length-dependent neuropathies,” Cornell said. “If the disease has progressed to a point where the much shorter nerves innervating the plantar flexors high up in the calf are affected, one will need a brace design to manage plantar flexor weakness as well as dorsiflexor weakness.”

Among several noteworthy takeaways Cornell found in the study were data revealing reduced control of the tibia over the foot throughout stance phase due to plantar flexion weakness.

“This indicates that it is the soleus controlling knee flexion throughout stance, not the quadriceps,” Cornell said. “This reinforces the need for ground reaction for these patients—an AFO to provide ground reaction for heel rise and proprioception for loss of balance. However, excessive crouch posture in static standing may be associated with proprioceptive loss and a strategy for improved balance.”

Misener noted: “I was a little disappointed on when they suggested bracing should be used—only after the patient presents with pain—and think they missed the mark. A ROM loss would likely not occur unless there was also a muscle imbalance.”

He said it’s critical for orthotists to be aware of gait abnormalities and address them appropriately with bracing—whether it’s as simple as a foot insert or a device with more aggressive muscular control, such as a fixed AFO to limit the amount of ankle dorsiflexion in stance, provide stability to the foot, and assist movement of the ground reaction force in the front of the knee during stance. 

Keith Loria is a freelance writer in northern Virginia.

Source: Wojciechowski E, Sman A, Cornett K, et al. Gait patterns of children and adolescents with Charcot-Marie-Tooth disease. *Gait Posture* 2017;56:89-94.



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## Update: Orthotic care and physical therapy for DMD

Lower extremity interventions can help boys with Duchenne muscular dystrophy (DMD) stay ambulatory for years—and improve outcomes in the condition’s nonambulatory phase. Devices can address contractures and other issues, while stretching programs are key to maintaining flexibility.

By Hank Black

Gene mutations in DMD short-circuit production or function of dystrophin, a protein that keeps muscle cells intact,<sup>1</sup> and the disease is marked by unrelenting muscle-fiber degeneration and progressive muscle weakness.<sup>2</sup> For this reason, lower extremity practitioners remain central to efforts to manage this disorder of X-linked recessive inheritance that affects 1 in 3600 boys (rarely, girls).<sup>3</sup> The emergence of progressive contracture and gradual loss of functional muscle are indications for orthotic use and the beginning of physical therapy.<sup>4</sup>

“Every provider’s expertise is directed at getting patients with DMD up and moving for as long as possible,” according to Kirk Ferris, CPO, director of prosthetics and orthotics at Mary Free Bed Rehabilitation Hospital in Grand Rapids, MI.

Diagnosis usually comes as children exhibit clumsiness and other motor signs of muscle weakness. Gowers’ sign—having to “walk” hands up the thighs to achieve an erect position—is due to weakness in the pelvic girdle and lower extremities and is a cardinal alert to the disorder.

Lower extremity interventions are essential to maintaining patient’s ambulatory phase. As muscles deteriorate, most children with DMD who do not receive corticosteroid treatment will need a wheelchair by age 12 years; with treatment, children may walk functionally many years longer.<sup>5</sup>

Orthotic intervention often begins when tightening muscles cause contractures and range-of-motion loss in the ankle, leading children to toe walk.<sup>6,7</sup> Toe walking is a characteristic gait in this population, one that a recent study suggested is an adaptive, “voluntary” mechanism meant to increase stability, rather than pathology.<sup>8</sup> Typically, a stretching program is initiated, with night splints and ankle foot orthoses (AFOs) prescribed to maintain gains from stretching.

Sara Rubinstein, CO, LO, TRS, orthotist and rehabilitation specialist at the Ann & Robert H. Lurie Children’s Hospital of Chicago, noted, “If the condition is apparent in infancy from genetic diagnosis with a known family history, then we have the chance to get involved



Jennifer Wallace, PT, helps this boy with DMD stretch his gastrocnemius and Achilles tendon. (Photo courtesy the Duchenne Therapy Network.)

“Every [lower extremity] provider’s expertise is directed at getting patients with Duchenne’s up and moving for as long as possible.” —Kirk Ferris, CPO

Continued from page 9

when the child first develops his nighttime routine. The helpful trick is to get him to tolerate nighttime bracing before contractures are present and as early as possible to create normalcy in the routine of positional bracing.”

Adherence is an issue. Although experts recommend using night splints in the nonambulatory phase, parents reported stopping use of night splints at a mean age of 10.3 years, after a mean duration of use of 2.9 years.<sup>9</sup>

## Orthoses: Day vs night use

Ciafaloni and colleagues found that, on average, 2.5 years pass between the appearance of the first signs and symptoms of DMD and a DMD diagnosis based on muscle biopsy or a DNA test. The average age at diagnosis for DMD is 5 years.<sup>10</sup> Because of this diagnostic delay, many children with DMD are initially misdiagnosed and may present to specialized clinics already walking in an orthosis. But daytime, functional AFOs for DMD are controversial, and most practitioners limit the devices to nighttime use.

Townsend et al found daytime AFO use decreases ambulatory function and increases incidence of falls.<sup>11</sup> That study has been criticized for using a dynamic, rather than an articulated, AFO and for not including kinematic assessments.

Sandra B. Smith, MS, PT, director of rehabilitation services at Shriners’ Hospital for Children—Tampa, in Florida, said: “Daytime AFOs are not usually tolerated due to the need for patients to adjust their center of gravity forward to balance, with increased plantar flexion at the ankle and increased lumbar lordosis, which occurs due to quad and gluteal muscle weakening.”

One recent small study of kinematics and kinetic gait assessment found potential benefit for daytime AFO<sup>12</sup> use by children with DMD. Lead author Ana Claudia Mattiello-Sverzut, PT, MSc, PhD, said her group recommends daytime wear when less than 5° of dorsiflexion is present. The results indicate that daytime ambulation with an AFO minimized the biomechanical compensations typical of DMD.

“In addition, our recent unpublished study showed daytime AFO use decreases the incidence of falls and is well accepted by patients with DMD if they choose to wear it,” said Mattiello-Sverzut, professor of pediatric neurology at the University of São Paulo, Brazil.

Rubinstein noted the published study showed improvements in the 10-m walk test. “My experience, however, is this may not be the case after a full day in the device,” she said. The São Paulo group did call for further study into whether their result would hold up over time.

“Daytime AFOs may be used as resting splints for ambulatory patients as a supplement for those who are not able to tolerate the AFOs the recommended six hours at night,” according to Jennifer Wallace, PT, owner of Duchenne Therapy Network and education director for the nonprofit CureDuchenne, both in Los Angeles, CA.

Leslie Vogel, PT, of Seattle Children’s Hospital in Washington, pointed out that older nonambulatory patients are often allowed to choose when to wear devices. “They often choose daytime because by then it’s harder to reposition themselves in bed with an orthosis,” she said. Children with DMD often are on respiratory support at night, making repositioning more difficult.

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"At night, the patient might get an articulated AFO with strapping to put a dynamic stretch to the ankle, but we use a solid AFO if they have plantar flexion contractions," Vogel said. "In the non-ambulatory stage of DMD, if the boy has a plantar flexion contracture, we may have him in ankle resting splints to prevent inversion and equinovarus."

Rubinstein, however, prefers not to use strapping. "Our patients say strapping is cumbersome when caught in blankets and often wakes kids up," she said. "Fewer moving parts with regular follow-up for necessary adjustments is ideal."

## Issues at the knee and hip

Another orthotic issue is how to address knee and hip flexion tightness. Rubinstein said, "We may tackle this problem with a knee immobilizer in conjunction with a positional AFO, but tolerance may be a challenge. More research is needed, but for now our bread-and-butter protocol is to use positioning and stretching programs with physical therapists to tackle the hip and knee tightness, with AFOs addressing the tendency for equinovarus contractures."

Most orthoses for children with DMD are custom-made to accommodate the fibrotic nature of calf muscles in this population, Rubinstein said. "We are exploring a static, progressive, solid design that we can adjust as kids change throughout the year. If we can adjust the angle and keep the child comfortable, he ultimately will wear the braces longer."

Rubinstein said, "Ideally, these boys will obtain a power wheelchair that accommodates standing. And if a KAFO [knee ankle foot orthosis] is considered, we do a trial period in knee immobilizers with PTs if a kid is highly motivated and starting to have increased

falls, and as long as his ankle, knee, and hip joints are not too tight. If we feel he is a good candidate for therapeutic use of KAFOs in a stander, I'll do a custom set with ground-reaction AFO portions."

Design and construction of a KAFO, orthotists said, require great precision to avoid disrupting knee mechanics and affecting the activity of more proximal muscles.

"Because the muscles above the knee are affected before those below the knee, and weakness proceeds from proximal to distal, a poorly aligned AFO can create abnormal flexion moments at the knee and overwork the quadriceps," according to Thomas V. DiBello, CO, FAAOP, director of clinical and scientific affairs at the Hanger Clinic in Houston, TX. "It's a very delicate orthotic process that requires practitioners who are highly experienced in tuning and alignment. Functional orthoses such as this need to be designed carefully. Their use is controversial."

Rubinstein added: "Tuning is crucial. It's important to have an alignment that accommodates any contractures but allows the child to have a forward center of mass to balance over their base of support with weak hip extensors."

Eric Weber, LCPO, at the Hanger Clinic in Seattle, WA, said, "KAFOs can be a useful treatment following surgical tendon release or lengthening. While in a standing frame or other safe condition, KAFOs provide a suitable base of support for proximal lower limb weakness."

Weight gain, a frequent side effect of corticosteroid use, makes ambulation more difficult, and a nutritionist or dietitian is now included in multidisciplinary clinics. In a healthy child, weight gain would be countered with an exercise program. However, children

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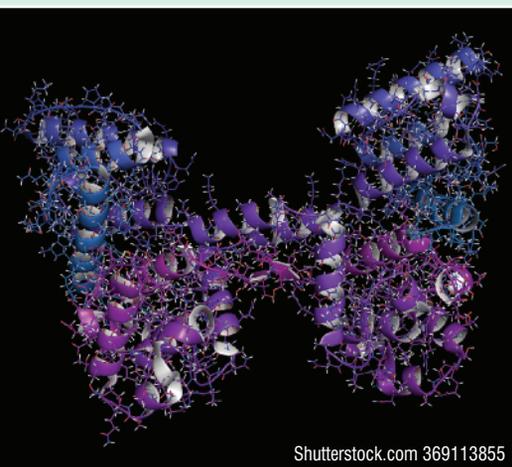
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## DMD medications

Much attention now is on the advent of molecular-based medications targeted at the underlying genetic defect of Duchenne muscular dystrophy (DMD) as well as on steroidal and nonsteroidal pharmaceuticals to control deterioration of the abnormal muscle.

Despite their side effects, prednisone and other corticosteroids add years of greater strength,

ambulation, and survival to a population whose life was once limited to their early twenties.<sup>16</sup> A 2014 study found steroid usage the most significant predictor of improved wheelchair-free survival. Now some patients are living into their fourth decade.<sup>17</sup>



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Molecular-based drugs enable production of the muscle protein dystrophin (represented above), which in DMD is blocked by gene mutations.

“There’s a lot going on that’s exciting,” according to Michael D. Sussman, MD, a pediatric orthopedic surgeon in Oregon at Shriners Hospital for Children—Portland. “Until corticosteroid treatment became accepted practice nearly 20 years ago we had nothing to really change the course of the condition, and now we have the first approval of molecular-based pharmaceuticals specific to Duchenne,” he said.

Molecular therapies target different genetic defects and include gene transfer or gene editing techniques. Eteplirsen, for example, the only Food and Drug Administration-approved medication for DMD, delivers through intravenous infusion an exon-skipping molecular “patch” that masks exon abnormalities and allows dystrophin production. It increases skeletal muscle but is only indicated for the 13% or so of children with DMD who have a confirmed mutation of the dystrophin gene amenable to exon 51 skipping.<sup>18</sup>

Ataluren is an oral “protein restoration therapy” for treatment of DMD caused by a nonsense mutation (about 13% of DMD patients). It’s approved for use in DMD in the EU and has been granted orphan drug status in the US. It overcomes the mutation by forcing cells to ignore a premature stop signal, enabling the production of the full-length, functional protein.<sup>19</sup>

References are available at [lerpediatrics.com](http://lerpediatrics.com).

with DMD must not overdo it, experts say, or they will suffer additional cell damage and muscle deterioration.

Smith said, “The therapist must monitor subjective signs of fatigue and continually reassess objective changes in function and symptoms during treatment sessions. Eccentric or high-resistance exercises should be avoided.”<sup>13</sup> She said Shriners-Tampa recently opened a multidisciplinary clinic for children with DMD and other neuromuscular disorders. Such teams of professionals are becoming more common at major centers as subspecialties’ diagnostic and management options broaden.

Vogel recommends starting powered mobility earlier, before total loss of ambulation occurs. “The boys want to keep up with their peers but shouldn’t be oversteering their muscles. We tell families that if the boy’s legs are sore at night or the following day, they’ve overdone it and need to decrease either duration or intensity of exercise.”

In fact, Vogel said, “Repetitive exercises such as leg lifts or sit-ups stress the same muscles. We view exercise more as simply activity. If they’re playing with peers, using a tricycle or bike with training wheels, or simply moving about, that’s about all they need. In addition, such activity provides more self-esteem and self-satisfaction than traditional exercise.”

Wallace said even a daily routine can become too stressful for boys with DMD. “Energy conservation becomes really important, especially for those aged seven to twelve years,” she said. “We want them to walk when they’re safely able to, but to use an electric scooter for those times they need to conserve energy to not deteriorate too quickly.” Changes may include avoidance of hills, for example, or a school schedule with classes closer together.

## Stretching, surgery, and other interventions

Stretching programs are essential for maintaining the flexibility of the hip flexors, iliotibial band, hamstrings, and plantar flexors. This allows boys to remain upright longer and to later be positioned comfortably in a wheelchair, Vogel said. Parents are vital to success of stretching, which should be performed six times a week, Wallace added.

When the child is aged 2 to 4 years, parents should start stretching the heel cords and hamstrings, Vogel said. “As they get into school, a stretch for hip flexors and iliotibial bands are added, and some kids are able to do their own stretches,” she said.

Serial casting was once a treatment for later-stage DMD to help maintain heel-cord stretches, but now has gained favor as an earlier treatment, Wallace said.<sup>14</sup>

Guidance published in the March 2018 issue of *Lancet-Neurology* by a multidisciplinary group of experts recommended foot surgery to improve varus positioning and Achilles tendon surgery to improve dorsiflexion.<sup>15</sup>

Achilles lengthening may be employed in carefully selected patients—that is, the 10% to 15% whose ankles experience a contracture of approximately 30° and whose proximal muscles are still relatively strong, said Michael D. Sussman, MD, pediatric orthopedic surgeon in Oregon at Shriners Hospital for Children—Portland. Children with DMD, he added, are prone to falls and fractures. “These must be treated very aggressively with good fixation,” Sussman said.

Standing support has become an important part of DMD care, including postsurgically. Wallace and Vogel said that use of a standing frame should be started while the child can still take a few steps—an example of interventions now used earlier than in the

past. "Standing frames provide stability to stay upright for home-work, standing programs at school or therapy, as well as for improving respiratory care, and delaying the onset of scoliosis."

Aquatic therapy and antigravity treadmills are heavily favored by physical therapists for children with DMD because they allow for more low- or no-impact exercise and are particularly useful for post-surgical rehabilitation, according to physical therapists interviewed for this article.

Overhead harness treadmills have fallen from favor, Wallace said, largely because the patient's weight falls on harness straps that go



Use of a standing frame should begin when the child can still take a few steps. (Photo courtesy of the Duchenne Therapy Network.)

through the groin area. One antigravity treadmill uses differential air pressure technology to provide lifting forces to reduce impact.

As Wallace conducts DMD educational sessions for other physical therapists around the country, she has noticed many regional differences in management. "My expectation is that these differences will largely disappear as new outcome measures and treatments are validated through clinical trials and as awareness of the progress made in Duchenne is raised through journals, conferences, and standards of care," she said. 

Hank Black is a freelance medical writer based in Birmingham, AL.

References are available at [lerpediatrics.com](http://lerpediatrics.com).

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Product Manual

## Media, toys, and games for kids with disabilities

Children use toys and media characters to spark their imaginations and cast themselves as the star of their own stories. Through play, they connect with other kids and dream about their future. Options are few, however, for kids who wear a device or otherwise look “different” to see themselves reflected during play.

By Jill R. Dorson

Melissa Shang has Charcot-Marie-Tooth (CMT) disease and spends much of her time in a wheelchair, though she can stand and walk short distances with her ankle foot orthoses (AFOs). As far as the 14-year-old is concerned, however, her physical ailments don't make her different from an average teenager.

She'd like to see the media and toy industries agree, and give her more characters—on television, in the movies, in books—and dolls that look more like her.

“I've read a few books that showed what living with a disability was like pretty well,” Shang said. “In the ‘Arlene on the Scene’ series, the main character has Charcot-Marie-Tooth, just like me. She uses leg braces, and in middle school she felt very self-conscious about herself and wanted to be like everyone else, but she finally just accepted herself.”

Shang has already done more than accept herself. She's busy trying to get other people to accept kids like her or, better yet, understand them.

“A lot of people seem to think that I sit around all day and think about my disability and all the things I can't do,” said Shang. “But I don't. I'm a teenager and I like talking about boys and crushes and I also get on my phone a little too much. I look forward to going to school and seeing my friends. I don't let my disability define me.”

In fact, Shang, who is in eighth grade and lives in Newton, MA, has been making her own grassroots effort to get the world at-large to understand better what it's like to have a disability. But she's finding what orthotists, physical therapists, and other lower extremity practitioners already know: there are few books, games, films, television shows, or toys that represent children with a lower extremity disability. When most professionals in the field are asked what's out there for kids, they're stumped.

Gordon Ruder, BSc, MSc, CO(c), FCBC, a Canadian orthotist who owns Boundless Biomechanical Bracing and is the program director at George Brown College's Department of Prosthetics and



Melissa Shang, who has CMT, petitioned American Girl to make a “Girl of the Year” doll with a disability. (Photo courtesy of Melissa Shang.)

Entertainment that either reflects the disabled community or is adapted to include it is critical to the mental well-being and self-esteem of kids with a lower extremity disability, according to practitioners.



Melissa and her sister Eva Shang (left) wrote "Mia Lee Is Wheeling Through Middle School," which features a strong main character who wears AFOs and is in a wheelchair. (Photo courtesy of Melissa Shang.)

Orthotics in Toronto, said there's not a lot available for practitioners to give to kids who are getting their first device.

"If it's going to come from anywhere, it's going to come from a patient-centered group," he said. "And usually in the form of a pamphlet or on their web page versus something children can connect with."

This anecdotal evidence is backed by the book *Disability and Popular Culture: Focusing Passion, Creating Community and Expressing Defiance*, by Kate Ellis, PhD, a senior research fellow in the Internet Studies Department at Curtin University, Perth, Australia.

"There is a distinct lack of research into disability toys from a cultural studies perspective," Ellis writes. "Indeed, when it comes to disability and toys, the majority of academic discussion centres on developmental psychology, physiotherapy and the construction of special toys for children with 'special needs.' In short, the discussion is entirely medicalized."

Despite limited resources, professionals say entertainment that either reflects the disabled community or is adapted to include it is critical to the mental well-being and self-esteem of children with a lower extremity disability.



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## Seeing a person—not a disability

"I think the more society is exposed, the more these characters are humanized," said Dana Dempsey MS, CTRS, the director of the Therapeutic Recreation Department at Texas Scottish Rite Hospital in Dallas. "There's a certain amount of wanting to empathize as well as wanting to support and advocate once you know the character. What happens in society is that those [children] are somewhat separated and [because of that], being disabled becomes a term more than a face or personality."

Board and table games, said Dempsey, are the obvious entertainment options that make things "even-steven"—meaning that all can participate. Books and television would seem the same, but as both Shang and Dempsey point out, there are many examples of disabled characters as "sidekicks," but few as protagonists.

Shang in 2014 started a petition asking the American Girl doll company to create a "Girl of the Year" with a disability. The company does have wheelchairs and two kinds of crutches available on their website as accessories but has not made a key character who is wheelchair-bound or wears AFOs. The 2012 "Girl of the Year" doll, McKenna, did have a tutor named Josie, who was in a wheelchair, but Josie wasn't the main character.

"Her job was to make the main character look better," Shang said. "That's not what people with disabilities are all about. The 'Girl of the Year' is my favorite part [of the American Girl brand], they make a doll [with a story] that will go out and conquer something. Through them, I was able to put myself into able-bodied things, like dancing, gymnastics, or horseback riding, but I've been disappointed because I was tired of seeing characters I couldn't relate to."

Shang's online petition has netted more than 140,000 signatures in support of an American Girl doll with a visible disability. Her plea was moving and direct:

*"Being a disabled girl is hard. CMT prevents me from activities like running and ice-skating, and all the stuff that other girls take for granted. For once, I don't want to be invisible or a side character that the main American Girl has to help: I want other girls to know what it's like to be me, through a disabled American Girl's story."*

The company contacted Shang and thanked her for her support but has not yet made a doll whose life story includes living with a disability.

## Entertainment options

Dempsey, as well as other professionals in the industry, are somewhat stumped when it comes to finding books, games, films, or anything else that represent disabled kids as more mainstream.

Toys "R" Us has a section on its website featuring toys for "Differently-Abled Kids," while FatBrain Toys has a section for kids with special needs; yet, neither offers dolls or toys that represent the physically disabled population. Most are geared toward children with a learning or mental disability.

When it comes to media, gaming, or literature, the entertainment options for kids with a disability widen a bit. Dempsey pointed out that many video games are perfect for kids with a disability: Whether it's a dancing game or air guitar or even Mario Bros., these kinds of games allow disabled kids to participate on a leveled playing field.

"The kids can be with each other and be engaged together," Dempsey said.

*Continued on page 18*

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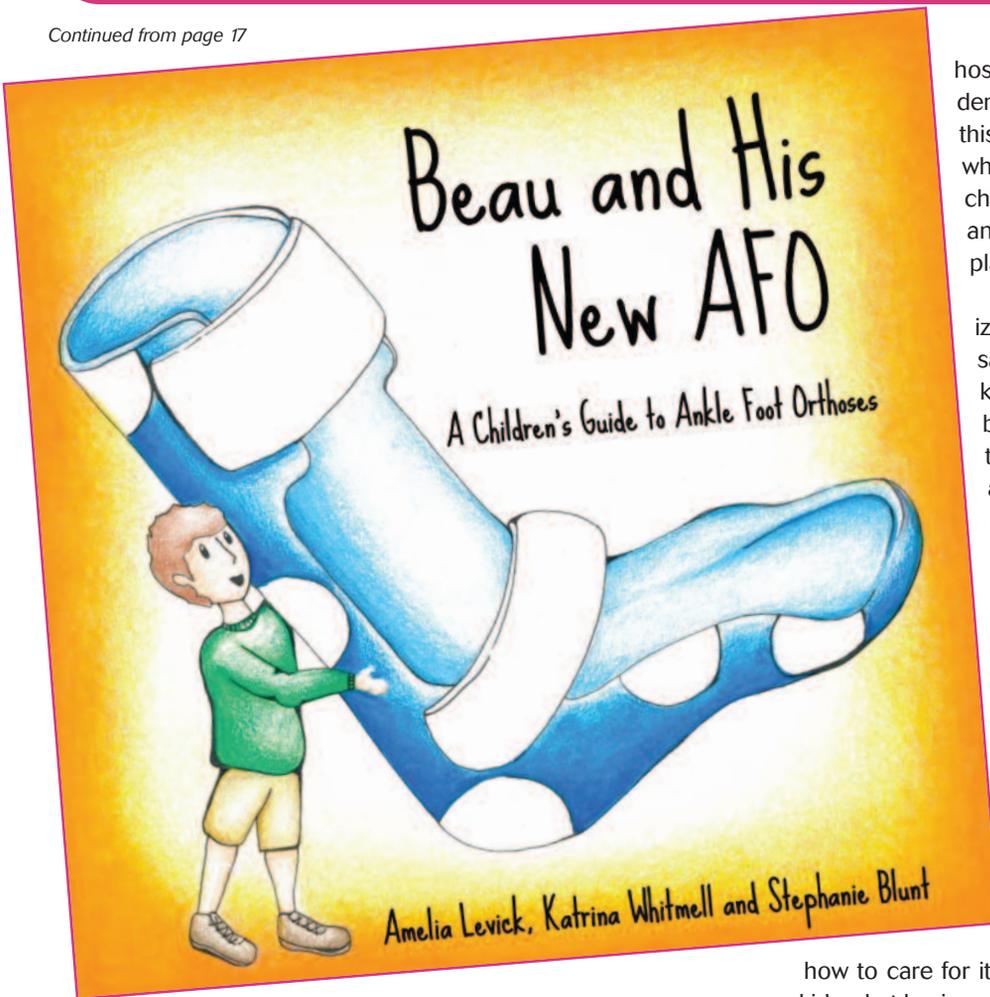
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A Canadian health system will publish "Beau and His New AFO" later this year. (Image courtesy of Stephanie Blunt.)

Ruder said he and his colleagues mostly have given pamphlets to patients or directed them to a special interest group for support. That's about to change: Ruder recently oversaw a student project at George Brown that led to the soon-to-be-published storybook, "Beau and His New AFO."

When Stephanie Blunt, an orthotic technician who graduated from George Brown College and is currently on pregnancy leave, was a student in Ruder's program, she and two of her peers, Amelia Levick and Katrina Whitmell, decided to write a book for kids with AFOs.

Its authors hope their book helps "normalize what it means to have an AFO." (Image courtesy of Stephanie Blunt.)

Hamilton Health Sciences, one of the largest hospital systems in Canada, partnered with the students, and will publish "Beau and His New AFO" later this year. Hamilton Health will distribute the book, which is geared toward teaching elementary-aged children about their device, to patients in its system and will create an audiovisual version that can be played inside patients' rooms.

"We wanted to give kids the ability to normalize what it means to have an AFO or device," Blunt said from her home near Toronto. "They never see kids with an AFO or in a wheelchair on TV or in a book. So, we wanted them to be able to say, 'Hey, there's kids just like me who wear devices.' There are two characters, one who is ambulatory and one who is in a wheelchair. That's important because not every AFO user can walk."

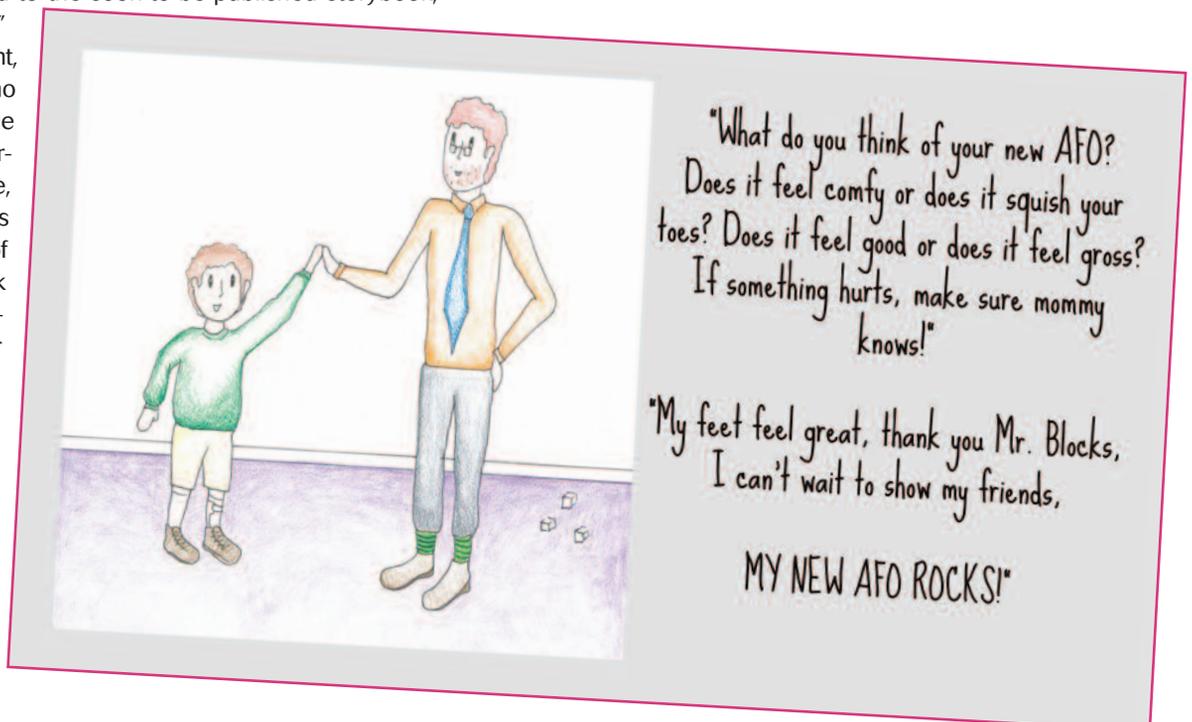
During her training, Blunt recognized that kids with a disability wanted to "connect with their toys and dolls" just like the average kid. She even worked with a child who asked if AFOs could be made for her American Girl doll.

"It's unfortunate that they can't pick a wheelchair or an AFO for their toys," Blunt said, "It makes them feel outcasted."

Although her book is more of an educational tool than a mainstream storybook, it is designed not only to help kids understand what they can and can't do with an AFO and

how to care for it, but to give them a tool for explaining to other kids what having an AFO means. Blunt hopes children will take it to show-and-tell and begin to educate their classmates about the device they use.

Shang has also written a book. "Mia Lee Is Wheeling Through Middle School" features a main character who wears AFOs and is in a wheelchair. The idea behind the book isn't so much to educate those around her but to create a strong main character whose



disability doesn't define her. The book was published in 2016 and is available through Amazon.

"Mia Lee is a star motion-picture maker and is trying to become the video club president. She meets a lot of people and makes a lot of new friends and she has to figure out if people are trying to be her friend because they feel sorry for her or because they really want to be friends," Shang said. "She grows from this experience."

## Learning by watching

For Dempsey, the recreational therapist, both Shang's and Blunt's books would fit right into her goal of helping to make her patients as independent as possible. When working with kids with disabilities, she has learned that, like other kids, they want to fit in and that they often learn by watching. Through Texas Shriners, she takes a group of her young patients with disabilities to specialty camps every year, where they can play with and learn from other kids with similar conditions. She believes the camps provide the opportunity for kids to see how others cope in like situations, but she's also seen that the development within this community is the same as it is anywhere else.

"Just like typically developing kids, they see each other on the playground and, well, how do you think kids learn how to use the monkey bars? They watch other kids," Dempsey said. "It's the same thing. If kids with disabilities can see how other kids are doing it, even if they can't do it the same way, they can take something away from it."

Patrick DeHeer, DPM, owner of greater Indianapolis' Hoosier Foot and Health and team podiatrist to the Indiana Pacers, said he has seen little he can use in his practice to either educate or enter-

tain kids who wear devices. Because of that, he directs kids and their parents to stay active in whatever way they can—running, jumping, swimming, or any other activity. Of particular note, DeHeer said he believes that, for kids with AFOs, baseball is a sport many can reasonably participate in.

"Getting an AFO into a baseball cleat isn't an easy thing, but I've had kids who have played baseball," he said. "It's a sport that kids with AFOs can be somewhat successful in."

He is enthusiastic about books like Blunt's.

"I don't have anything like [Beau and His New AFO]," DeHeer said. "But I think it's brilliant."

The author thinks so, too.

"This is a niche that is really untapped," Blunt said. "I think it has great potential for our population. And we made it generic on purpose, so we could adapt it to kids from another population, like kids with spina bifida or cerebral palsy."

For Shang, while putting disabled kids front and center is something of a passion, it is more of a hobby than a feature that defines her. But as a disabled person herself, she'd still like to see the larger world acknowledge her existence through characters—whether physical, like dolls, or in books, games, magazines, or films—who are more mainstream than disabled.

"I want people who don't have disabilities to have a better understanding of what it's like to live with a disability and what our lives are really like," Shang said. "And I want people with disabilities to know that they can succeed and to see that they are properly represented." 

Jill R. Dorson is a freelance writer in San Diego County, CA.

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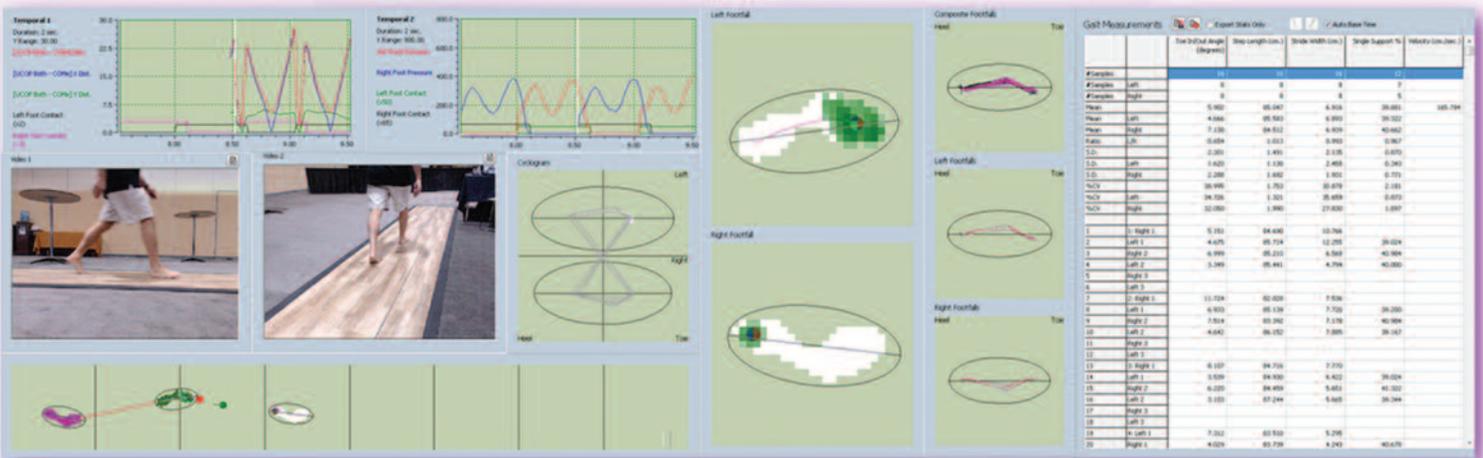
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