

Lower Extremity Review

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*Orthotic management of CMT:*

## DYNAMIC SOLUTIONS

*for active lifestyles*

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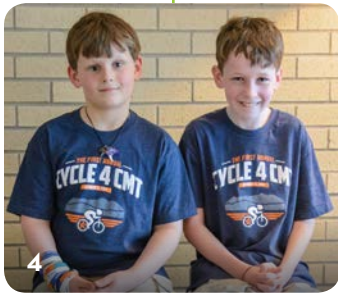
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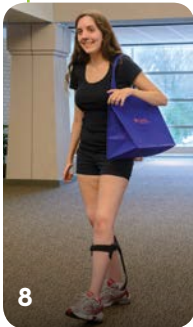
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## FROM THE EDITOR:

### *Meet your match*

For people with Charcot-Marie-Tooth (CMT) disease, it's not easy to find a healthcare professional who understands the disease and its complexities. Sometimes a patient has to visit multiple clinicians before finding one who is right for the job. But finding that perfect match—someone who knows what symptoms to look for and which interventions will work best—can literally change a patient's life, from one filled with frustrations to one focused on solutions.

The same can be said for finding the right orthotic device. Given the vast numbers of CMT subtypes, and the fact that even patients with the same subtype may have different symptoms, it's entirely possible that the first ankle foot orthosis (AFO) a patient tries may not be the best match for that person's symptoms, activity level, and lifestyle priorities. A patient may need to test-drive multiple AFOs before finding the one that's the best fit in all senses of the word.

But, like finding the right clinician, finding the right AFO can be life-changing. At the most basic level, an AFO can improve gait and balance. But those improvements lead to other benefits—greater confidence, a more active lifestyle, a more positive outlook.

That's what this special issue is all about. On one level, it's about matching the right AFO to the right patient. But really, it's about changing lives. In the pages that follow, you'll find the information you need to do both.

BY JORDANA BIEZE FOSTER,  
EDITOR





## Orthotic devices and CMT: Everyone deserves an aha! moment

This guide to orthotic devices for people with Charcot-Marie-Tooth (CMT) disease has been a dream of mine for a long time. I was diagnosed with CMT1A at the tender age of 18 months, but didn't learn about orthotic devices for CMT until 2007, when I started a support group in Atlanta. Up to that point, I thought ankle foot orthoses (AFOs) were for the aged and infirm—something that, at age 35, I didn't think I was.

When orthotist Sean McKale, CO, LO, came to my support group meeting, he immediately recognized in me the walk of someone with CMT. When Sean, who sits on the CMT Association (CMTA) Advisory Board, shared this with me, I was shocked. I had no difficulty walking—or so I thought! However, I listened closely as he talked about the benefits of in-shoe foot orthoses to correct alignment and prevent joint deterioration. My mother had just had a double knee replacement, necessitated by poor alignment caused by her CMT. OK, I thought, maybe I could use foot orthoses to prevent that type of joint deterioration.

As Sean went on to detail the benefits of AFOs, especially the dynamic AFOs, I got progressively more excited. Did I want to fall less? Yes. Did I want more energy throughout my day? Yes! Did I want to stop reduce my joint pain, especially in my knees, hips, and back? YES! OK, Sean, I thought, I'll try a pair of AFOs.

That was my aha! moment. The AFOs felt like very light-weight, stable boots with springs in the heels. They looked like shin guards, the kind used in soccer. They could be easily camouflaged under pants, with the right shoes. Yes, I thought, maybe I could use these from time to time.

Six years later, I wear them about 50% of the time. I love to hike in the woods, where my AFOs prevent trips and falls. I love to travel, and my AFOs prevent fatigue from long walks and rolling my ankles on cobblestones. Overall, my AFOs allow me to walk further, longer, and to have less pain and more energy at the end of an arduous day. I can do the things I love to do!

I have not become dependent upon my AFOs. I don't wear them when I know I won't walk far or when I would prefer to wear shorts or a dress. I don't wear them to exercise on the bicycle or treadmill, where I can hold on for balance. I have learned when I can go without them and when I can't. I have worked hard to maintain strength and muscle tone in my legs. My AFOs supplement my leg strength rather than replacing it, vastly improving my quality of life.

This guide is aimed at helping others to reach their aha! moments faster than I did.

Together with *Lower Extremity Review* and the extensive input of CMTA Advisory Board members Sean McKale and David Misener, CPO, we hope this publication will empower patients with CMT and practitioners to discuss different types of orthotic devices and make an informed decision about which type will be the best fit.

The CMTA's vision is a world without CMT. We will achieve this vision through our extensive and focused drug development research initiative, STAR (Strategy to Accelerate Research). We know that we will soon be successful in finding treatments to slow, stop, and reverse the progression of CMT.

Until then, we believe it is our responsibility to provide resources to people living with CMT about treatments that can improve quality of life, and, when indicated, those include in-shoe foot orthoses and AFOs.

I encourage you to visit our website, [cmtausa.org](http://cmtausa.org), for research updates, educational resources, to connect with others in the CMT community, or to invest in our vision of a world without CMT by making a donation. And please feel free to call us at 800/606-2682 or email us at [info@cmtausa.org](mailto:info@cmtausa.org).

Wishing you the best of health,

Susan

SUSAN RUEDIGER  
DIRECTOR OF DEVELOPMENT  
CHARCOT-MARIE-TOOTH ASSOCIATION



# Overview of CMT: Clinical challenges and opportunities

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*Early diagnosis and early intervention in people with Charcot-Marie-Tooth disease give clinicians and patients a valuable head start on keeping symptom progression in check.*

David Misener, CPO, understands Charcot-Marie-Tooth (CMT) disease firsthand because he has it himself, as does his son. Misener, who practices with Clinical Prosthetics and Orthotics in Albany, NY, said his case was difficult to diagnose at first because he and those in his family don't carry the genetic mutation usually associated with the disease.

Photo by Joshua Albanese  
([joshuaalbanese.com](http://joshuaalbanese.com))



*“Because CMT affects both motor and sensory nerves, patients can experience not only muscle weakening and imbalance, but also problems with proprioception.”*

“They first ran the test on CMT1A, because that’s the most common, but they didn’t find the error,” he told *LER*.

As it happens, his family carries a different genetic mutation, CMT1B, which has the same symptoms but originates in a different gene.

“If you’re not one of those with type one-A, they have to keep looking, and it can become expensive,” Misener said.

### CLOSING THE INFORMATION GAP

CMT, a sensorimotor neuropathy marked by progressive muscular atrophy, was first described in 1886 by physicians from France and England. The disease typically begins in the intrinsic muscles of the foot, then extends to the peroneus brevis and longus, the tibialis anterior, the extensor digitorum longus, and the extensor hallucis longus. It may eventually involve the hands, as well, and it’s even been associated with seemingly unrelated conditions such as scoliosis.<sup>1,2</sup> Roughly one in 2500 people in the US has CMT, according to the National Institute of Neurological Diseases and Stroke (NINDS), and men are affected more often than women, in approximately a 5:3 ratio.<sup>3</sup>

David Misener’s diagnostic challenges occurred 20 years ago, and since then researchers have continued to refine their understanding of the genetic mutations that cause CMT. Research has revealed ever-expanding subclasses of the disease, in fact, which is usually inherited in an autosomal dominant pattern (ie, if one parent has CMT, each child has a 50/50 chance of inheriting it). However, some CMT subtypes may be passed on in an X-linked, or recessive, pattern, and the disease may also arise due to spontaneous or de novo mutations.<sup>4</sup>

“The most common subtype, CMT1A, affects about half of CMT patients,” said Michael Shy, MD, a professor of neurology, pediatrics, and physiology at the University of Iowa’s Carver College of Medicine in Iowa City. “As of now, four subtypes account for ninety percent of genetically identifiable CMT: CMT1A, CMT1B, CMTX, and CMT2A.”

According to Shy, CMT1A is considered the classic phenotype.

“These patients are likely to have been slow runners and clumsy as children, and as they reach adulthood many will need foot orthoses or ankle-foot orthoses (AFOs) to help them ambulate,” he continued. “Only a small percentage will ever need a walker or a wheelchair.”

Other subtypes exhibit different characteristics, Shy said. For example, some CMT1B patients don’t begin to walk until two or three years of age and are severely affected in the first two decades of life, as is also the case with many CMT2A patients.

“It just depends on the particular mutation within the subtype,” he said. “It can range from severe early on to very mild, though as a rule the recessive types—such as CMT4—are often fairly severe.”

### PATHOPHYSIOLOGY

CMT pathophysiology has traditionally been classified as either predominantly demyelinating or axonal, but clinicians and researchers disagree about the relative importance of these categories with regard to manifestation and progression of the disease.<sup>2</sup> For example, although axonal degeneration is a predictor of disability—suggesting that axonal damage may be the root cause of the neuropathy—the mutations responsible for the different forms of CMT1 typically occur in myelin genes, and myelin disturbances lead to axonal damage. Further muddying the waters is that axonal damage can result in secondary demyelination.<sup>2</sup>

Genes cause CMT by disrupting particular proteins, Shy explained, and if those proteins form a constituent of myelin, the myelin will be damaged.

“You will see axonal damage in CMT1,” he said, “but the primary problem is the myelin.”

Some aspects of CMT remain perplexing, however.

“Many of the genes that cause CMT2—the axonal form of the disease—are expressed widely throughout the body, not just in neurons,” Shy said. “So why other organ systems aren’t affected is a mystery. It probably has something to do with the biology



Photo by Jordana Bieze Foster.



of the neurons, since they don't divide, and have long axons that have to be maintained throughout the person's lifetime."

Axon length likely explains why distal parts of the body, starting with the feet, are affected first. And, because CMT affects both motor and sensory nerves, patients can experience not only muscle weakening and imbalance, but also problems with proprioception, Shy said.

Some muscles weaken more quickly than others, moreover, which leads to imbalances between dorsiflexors and plantar flexors. These imbalances, in turn, contribute to contracture of the Achilles tendon and problems such as cavus feet, calcaneal inversion, forefoot adduction, and claw toes.<sup>5</sup> Other CMT-related problems include skinny calves, scoliosis, foot drop, "slapping" gait, foot numbness, muscle weakness, balance difficulties, and multiple non-neuropathic pain symptoms.<sup>2,4</sup>

## DIAGNOSTIC CHALLENGES

The heterogeneity of symptoms associated with CMT, and the subtlety with which they often present, may lead to delayed diagnosis in some patients.

"Many times people get diagnosed well after the age of onset," David Misener said. "Those with mild cases may not realize they have the disease until they're in their sixties and develop a drop foot."

Because more than 80 CMT subtypes have been identified, the broad range of severity and age of onset can also contribute to missed diagnoses.

"In many families with CMT, you sometimes reach a tipping point where something gets your attention, such as stumbling," Misener continued. "I think the disease probably progresses at an even rate, but certain events make you think it may have advanced more suddenly."

Michael Shy agreed.

"In CMT1A patients, the disease tends to develop slowly through the first two decades of life," he said. "Some CMT1B can present early, others don't present until adulthood. It depends on the particular mutation."

In making a diagnosis, Shy typically looks for length-dependent weakness and sensory loss, particularly in the hands and feet.

"Usually we look for absent ankle reflexes," he said. "In some cases, all the deep tendon reflexes can be absent, and then we use nerve conduction testing to determine whether or

*"Patients usually present with one or more of five key symptoms: poor balance, foot drop, lateral ankle instability, sensory loss, or painful calluses."*



Photo by  
Vincent Giordano  
Trinacria Photography  
(trinacriaphotography.com)

not it's primarily a problem with myelin or the axons."

Although Shy and other specialists are attuned to CMT, many clinicians are not. The result, he said, is that the initial diagnosis is sometimes made by a foot specialist rather than a primary care physician.

"I think podiatrists are often the first people to recognize that someone may have CMT, because they see pes cavus or other foot abnormalities," Shy said. "Length-dependent weakness and atrophy, and decreased deep-tendon reflexes are things that primary care physicians may pick up, though."

Ken Cornell, CO, who practices with Cornell Orthotics & Prosthetics in the Boston area, finds himself making the initial diagnosis less often than he used to.

"Twenty or twenty-five years ago I was getting a lot of these patients, and I'd call their doctors to discuss CMT, and they wouldn't even know what I was talking about," he said. "In the last few years that's started to change, I think partly due to outreach from the CMT Association."

Cornell said his patients usually present with one or more of five key symptoms: poor balance, foot drop, lateral ankle instability, sensory loss, or painful calluses. And, although nerve conduction studies have typically been considered definitive, increasing knowledge about the genetic basis of CMT has elevated demand for genetic testing.

## DISEASE PROGRESSION

Once the diagnosis of CMT is made, through either traditional means or genetic testing, clinicians and patients must monitor the disease's progression to determine the most appropriate interventions. As noted previously, the disparity in weakness between opposing muscles may ultimately lead not just to imbalances but to deformities.

"The longest nerves in the body are affected first," reiterated Ken Cornell. "In a man who's six feet tall, one single nerve cell going from the spine to the foot can be three feet long. That cell is affected first, and that in turn has effects on the muscles that extend the toes. But the muscles that flex the foot are higher up in the leg; they're shorter nerves, affected later, so you wind up with unopposed toe flexion. That's how you get claw toe deformity."

“The nerves are not feeding the muscles, so they start wasting away, and this creates muscle imbalances over specific joints,” added David Misener. “As one muscle pulls harder over that joint, it can affect the shape of the foot, which becomes cavus. Drop foot arises because the anterior muscle group weakens before you reach that tipping point in the calf muscle, and those anterior muscles lift the foot during swing phase and decelerate it at heel strike. These things start with the small intrinsic muscles in the feet, then affect the ankles, then the knees, and in some people, the hips.”

Cornell added that these disease traits make progression reasonably straightforward to predict.

“People are typically focused on the motor loss first, which usually manifests first with foot drop, then with everter paresis,” he explained. “The peroneus longus—the plantar flexor of the first ray—outlasts its antagonist, the anterior tibialis, so you wind up with a plantar flexed first ray and a tripod effect. This causes an inversion twist to the rearfoot, which sets you up for lateral ankle instability and forefoot adduction. All these muscle imbalances are progressive, and they set up the conditions for deformities.”

Michael Shy noted that certain types of nerves seem to develop problems more quickly than others, and it may not always have to do with nerve length.

“For reasons that are unclear, the nerves that cause dorsiflexion and eversion seem to get damaged earlier than those that cause plantar flexion and inversion, so you get a lot of anterior calf wasting,” he said.

With all the attention to motor nerve deficits, Shy added, CMT’s effect on sensory nerves shouldn’t be downplayed.

“Most CMT types affect both motor nerves and the large-fiber sensory peripheral nerves, which tell our brain where we are in space,” he said. “This can cause problems with balance. Some forms of CMT affect only motor nerves, others affect primarily sensory nerves, and why that difference exists isn’t completely understood yet.”

## INTERVENTIONS

Although CMT isn’t curable, interventions such as physical therapy, exercise, and orthotic device use can ameliorate its effects.

“We recommend stretching and moderate-intensity exercise for CMT patients,” said Katy Eichinger, DPT, CS, who practices in the neuromuscular division of the Department of Neurology at the University of Rochester in New York. “Patients should be evaluated by a physical therapist, who can guide them to a program that’s best for them. Assessing a person’s strengths, weaknesses, age, disease progression, and personal desires can make an individualized program much more successful.”

In terms of stretching, Eichinger emphasizes the ankle plantar flexors (a calf stretch, facing the wall with the leg extended backward and the heel on the ground, is a typical approach). For strengthening, she works with patients’ likes and dislikes to find something they can stick with.

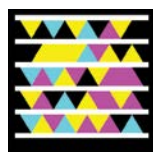
“You can optimize patients’ functional ability and make sure no weakness occurs due to disuse or aging,” she explained. “If someone doesn’t like gyms, they may be willing to do aquatic exercises or ride an exercise bike at home.”

Because CMT patients have distal weakness, physical therapy should focus exercise programs more proximally, according to Eichinger. This means an emphasis on the quadriceps, the hip muscles, and the core. It also may mean balance training, particular in those whose neuropathy forces them to rely more on their vision and vestibular systems to maintain an even keel.

## WHERE WE GO FROM HERE

With improved genetic sequencing techniques and increased understanding of proteins and myelination, researchers are moving closer to finding a cure for CMT. Today, the best treatments for CMT include regular exercise, maintaining a healthy body weight, and, when necessary, the use of orthotic devices. Approaches to orthotic management are discussed in detail in the article that follows.

CARY GRONER IS A FREELANCE WRITER IN THE SAN FRANCISCO BAY AREA.



References are available at [LERmagazine.com](http://LERmagazine.com), or by scanning the QR or tag codes at left.



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## Orthotic management: *A symptom-based, dynamic approach*

*Orthotic devices, tailored to each patient and adjusted for disease progression, can improve mobility by addressing gait impairments and maintaining stability.*

Orthotic management of the muscle imbalances associated with Charcot-Marie-Tooth (CMT) disease is a critical part of preventing or delaying later complications, which may include foot deformities and severely impaired gait. Because CMT is caused by genetic mutations that affect the nerves, such interventions don't affect the progression of the disease itself; they can, however, help ameliorate its worst manifestations.

Photo by Joshua Albanese ([joshuaalbanese.com](http://joshuaalbanese.com))



*“Orthotic interventions don’t affect the progression of the disease itself; they can, however, help ameliorate its worst manifestations.”*



Photo by Vincent Giordano  
Trinacria Photography (trinacriaphotography.com).

Clinicians wrestle with balancing correction and accommodation when prescribing orthotic devices for patients with CMT, and such decisions depend partly on the patient’s condition and wishes, as well as on the progression of the disease.<sup>1</sup>

What’s most important to patients isn’t always at the top of the list for practitioners, however.

“I ask my patients to prioritize,” said Ken Cornell, CO, who practices with Cornell Orthotics & Prosthetics in the Boston area. “Usually they complain about lateral ankle instability, painful calluses, foot drop, or claw toes. Only rarely do they mention balance problems, but I’ve found that, when you restore their balance, they suddenly realize what a big problem it was.”

Cornell agreed that orthotic intervention doesn’t address the underlying pathologies associated with CMT—an ankle foot orthosis (AFO) doesn’t give patients back their lost muscle strength or proprioception—but it can do other things.

“What’s challenging and exciting is restoring both static and dynamic balance by derotating the foot and trying to restore its alignment,” he said. “That changes the path of the center of pressure as it tracks over the foot, and that improves patients’ functional balance.”

According to Cornell, skilled orthotists can also address the deterioration that may lead to a “slapping” foot during the stance phase of gait.

“Particularly with dynamic carbon bracing, we can restore those three stance-phase rockers to normalize someone’s gait pattern,” he explained. “They have a heel strike, then controlled plantar flexion as the foot comes down to the ground. They’re getting dynamic resistance from the carbon so it’s giving them a ground reaction that tells them where their center of gravity is over their base. Then the tibia advances over the foot until it gets resistance in dorsiflexion, and then it raises the heel, which is the normal third rocker.”

Research supports the efficacy of AFOs for treating gait issues in CMT patients. One 2012 study from the UK, for example, noted that CMT patients often compensated for foot drop by increasing hip flexion during swing. In 14 individuals, three types of AFOs designed to address foot drop improved both proximal and distal leg control, decreased hip flexion amplitude during swing, and increased both ankle dorsiflexion and foot clearance.<sup>2</sup>

In a study noted in *LER* last year,<sup>3,4</sup> researchers fitted eight

CMT patients with custom carbon-fiber braces and reported that participants walked faster with the braces, particularly if they were relatively weak to begin with. As velocity increased, moreover, maximum joint moments during loading response shifted from the hip to the ankle and knee joints; the hip joint dominated during propulsion.

## CLINICAL EXPERIENCE

Overall, however, the relative paucity of research on orthotic treatment of CMT means that clinicians often develop approaches based on their own experience and that of their colleagues. Collaboration is often a key element of success.

“The literature as to when to use orthotics or AFOs, or when to perform surgery, is very poor,” said Michael Shy, MD, a professor of neurology, pediatrics, and physiology at the University of Iowa’s Carver College of Medicine in Iowa City. “It’s almost always based on a person’s clinical judgment. I’ve been fortunate enough to work with many talented orthotists who’ve all seen a lot of patients with CMT. They understand the foot structures and disease progression, and they’ve seen people respond over time. They’re partners in providing care, and they know more about their field than I do.”

Although genetic screening has led to the identification of at least 80 subtypes of CMT, when making treatment decisions orthotists are more concerned with how the condition presents.

“It’s not really diagnosis-specific, because as an orthotist you have to deal with the biomechanics that are present,” said Sean McKale, CO, LO, practice manager at Midwest Orthotic and Technology Center in Chicago. “People with the same type of CMT may be at different stages of progression, or just affected very differently, so you have to evaluate them as individuals and assess their symptoms to address their needs.”

One of the first factors McKale evaluates is a patient’s strength—or, more precisely, how much of it they’ve lost.

“Muscle weakness plays a big role in terms of decisions about level of support and the dynamics of the orthotic device,” he said.

Other concerns are the position of the foot, loss of range of motion (ROM), triplanar deformity, and proprioception.

“You use these clinical findings to help guide you, to show you the path to take with that patient,” McKale said.



Photo by Susan Ruediger.

McKale believes functional alignment correction is more important than accommodation in most cases.

"I think that with accommodation you end up allowing the foot to become more deformed," he said. "The exception is when there's been surgical fixation or fusions, which limit range of motion so that you can't gain functional corrections." In such cases McKale prefers a stable nondynamic device to maintain an aligned position.

In more typical cases, however, his goal is to balance the foot as well as possible to prevent further deformity.

"Someone with better proprioception can deal with increased dynamics," he said. "You have to vary the device you select, its stiffness and trim lines, based on the dynamic needs of the patient."

According to Geza Kogler, PhD, CO, director of the Clinical Biomechanics Laboratory in the School of Applied Physiology at the Georgia Institute of Technology in Atlanta, addressing common gait issues with orthotic devices can make important contributions to patients' quality of life.

"The ability for someone to lift their toe during swing phase has a dramatic impact on their ability to walk efficiently," Kogler said. "That's the primary benefit of the orthosis; a secondary benefit is that it slows down the foot during heel strike and stance phase, to help minimize foot slap. It also helps with stability during standing, and these things together have a profound impact on patients."

Kogler agrees with McKale that alignment should be maximized. In some cases, however, it isn't possible.

"Over time, with the loss of muscle, there can be contractures of the calf musculature, and that can lead to permanent deformity," Kogler said. "That can change the alignment so that you can't get the foot into a neutral position during standing, and in those cases you have to accommodate that alignment. You might have to put a lift under the heel to reach neutral, for example."

## ALLOWING THE MUSCLES TO WORK

According to David Misener, CPO, who practices with Clinical Prosthetics and Orthotics in Albany, NY, orthotists typically

strive to find the best balance between correction and accommodation. Misener has a deeper experience of CMT than most clinicians because the disease runs in his family and he has it himself.

"You have to be careful not to overbrace someone," he said. "The muscles need to work. Orthoses do need to support and align the body, though, and that can be as simple as an in-shoe orthosis. You can start with a very low profile device, inside the shoe, and then as the disease progresses, start working your way up the chain."

In his practice, however, Misener frequently sees patients who haven't been diagnosed early enough to allow for more minimal interventions.

"Ideally you want preventive orthotic management to maintain range of motion around specific joints," he said. "To balance correction and accommodation, I think we have to be more proactive in getting functional corrections of alignment around joints, because bracing becomes exponentially more difficult when more range of motion is lost."

According to Misener, patients should never be made physically uncomfortable by their devices, but other factors can also affect compliance. His experience is that CMT patients resist wearing corrective devices because they want to feel as "normal" as possible. This tends to backfire, however, as they start to lose range of motion.

"Ideally, you'll get a diagnosis early and start with simple corrective orthoses," Misener said. "Then, if you're starting to get muscle weakness around the ankle, or foot drop, a carbon brace will still allow range of motion and allow the muscles to work without over-supporting them. I think having some fatigue throughout the day is good, because it means you're exercising and strengthening your muscles."



Photo by Vincent Giordano  
Trinacria Photography  
(trinacriaphotography.com).



Maintaining range of motion is paramount as CMT progresses, Misener said.

“The key is to get the foot in subtalar neutral so you’re really stretching what you need to be stretching,” he said. “Night splinting [for Achilles tightness] can help, particularly with kids. They don’t want to wear braces, but if they do they’ll often gain range of motion.”

## DISEASE PROGRESSION

As Misener evaluates disease progression in his patients, he tries to address problems with the least drastic intervention possible.

“I think dynamic braces are a solid way to go,” he said. “Carbon systems are great because they’re lightweight and reduce fatigue, but you have to get the alignment correct. I like to be a minimalist whenever possible, but if I have to build something to overpower a muscle imbalance, then I have to go stronger. But I try to go stronger in a more dynamic way; you’d like to have the individual control their body as much as they can.”

Geza Kogler agreed that it’s crucial for the clinician to adapt to disease progression. In his experience, orthotic treatment and stretching should work together.

“A patient may go for years wearing an AFO that sets an alignment, but if they don’t keep up their stretching routine, they can end up with significant foot and ankle deformities that can complicate the fit of that AFO,” he said.

Kogler sees patterns in adjusting orthotic devices to disease progression.

“Early on, if someone has drop foot, a dorsiflexion-assist AFO might be enough, so the ankle can still move. But with time, if they develop more of a contracture, that assist will be overcome by the patient’s calf muscles, and they’ll have to switch to a solid-ankle AFO. But if they’ve stuck to a good stretching regimen, they may be able to avoid that step. It can’t always be avoided, but it can be delayed,” he said.

Ken Cornell agreed that a primary goal of orthotic intervention is to prevent deformity as CMT progresses, but that strategies depend on the individual patient.

“There is no one CMT brace, because everyone has a different level of gadget tolerance,” Cornell said. “You really have to make the patient part of the plan. If I apply too much pressure, to the point that the brace is uncomfortable and breaking

down skin, no one is going to wear that. You have to understand the patient’s priorities.”

## THE ROLE OF PHYSICAL THERAPY

As noted, physicians and orthotists play a big part in determining how patients respond to orthotic interventions. But physical therapists often play an important role, too, particularly because they often work with patients who have just received an orthotic device.

“Walking is an automatic task for most of us, but when people develop weakness or sensory changes, it requires much more cognitive attention,” said Katy Eichinger, DPT, CS, who practices in the neuromuscular division of the Department of Neurology at the University of Rochester in New York. “In slowly progressive conditions such as CMT, the body adapts and gait changes over time. We can be instrumental in helping patients relearn walking, by using different strategies and gait-training techniques to accommodate assistive or orthotic devices.”

For example, as part of these overall goals, physical therapists can help patients compensate for sensory loss, according to Eichinger. As clinicians have noted above, however, the success of such strategies has partly to do with getting the patient on board.

“Some people don’t like to use a cane, but they’ll use a hiking pole,” Eichinger said. “That may be all they need; you can see their gait improve because the pole provides increased sensory input about their position in space. It’s just a matter of teaching them to accommodate assistive and orthotic devices to maximize their functional abilities and gait techniques so they’re as energy efficient as possible.”

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“Physical therapists often play an important role in the management of CMT, particularly with patients who have just received an orthotic device.”

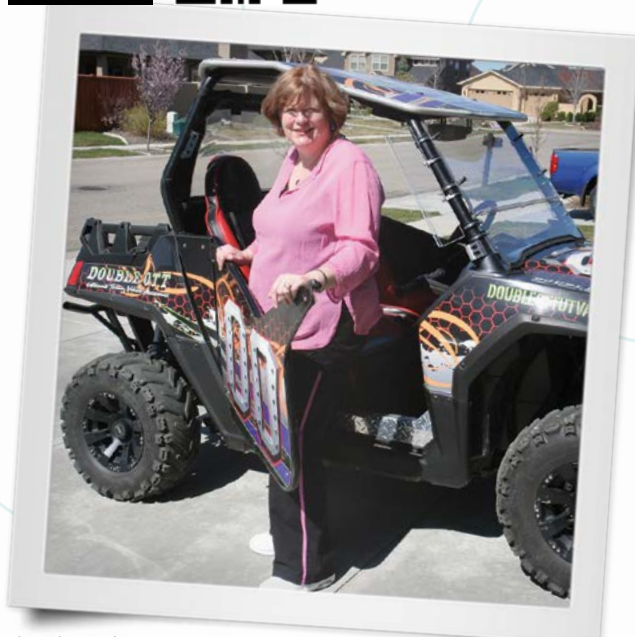


Photo by Jordana Bieze Foster

**Diane Covington:**

*AFOs positively impact every aspect of her dynamic life*

STORY AND PHOTOS BY JORDANA BIEZE FOSTER



It's a happy coincidence that Diane Covington lives just a few miles from a New Balance store, for two reasons. The brand's sporty and comfort styles are perfectly suited for Covington's life on the go. And they come in widths that can accommodate the ankle foot orthoses (AFOs) that make her active lifestyle possible.



Covington, 59, was diagnosed with Charcot-Marie-Tooth (CMT) disease 30 years ago, after coworkers' comments about her limping gait prompted her to see a neurologist. Later, when blood testing for CMT had become more widely available, her subtype was identified as CMT2A—and, in retrospect, she believes her father and paternal grandmother likely had CMT as well. The oldest of Covington's three adult children has also been clinically diagnosed with CMT.

Covington's symptoms include impaired balance, muscle weakness, and diminished sensation in her extremities. But, like many people with CMT, Covington was reluctant to try AFOs until her symptoms had deteriorated to the point at which she

could not stand unassisted and became fatigued after 15 minutes of shopping at the mall. That was nine years ago. Now, she wears her Custom Composite AFOs literally from when she wakes up until she goes to sleep (except when bathing), and she wishes she had embraced the idea earlier.

"If I had to say anything to people with CMT about AFOs, I would say, 'Don't wait,'" Covington said. "We think it's going to be this devastating experience, but in fact it's just the opposite. There is nothing about my life that my AFOs haven't impacted in a positive way. I think they're just the best."

The stability and support provided by the AFOs allows Covington to pursue a number of outdoor activities near her home outside of Boise, ID, including Nordic walking, riding a recumbent tricycle, and exploring the local landscape in an ultra-terrain vehicle with her fiancé, Mike Steiner. But she says some of the smaller achievements have made the biggest difference.

"I can now stand up by myself in the middle of a room," Covington said. "That doesn't sound like a big feat, but before I would have had to hold onto something in order to stand. I really would not be able to leave the house safely without them."

To be sure, finding the right pair of AFOs has been a process. Covington estimates she has tried 10 different pairs of AFOs and seen at least three orthotists, and not just because she and Steiner relocated to Idaho from Colorado last year. Some designs were uncomfortable, some were not durable enough, and some were too wide to fit in any conventional footwear. It took about six years to finally find her AFO soulmates.

"I would also tell people with CMT to not be shy about changing orthotists if the two of you are not on the same wavelength," Covington said. "Find someone who understands CMT, because not everyone does."

Over time, Covington has become more comfortable disclosing her condition to others—something that was particularly challenging when she reentered the dating world several years ago as a divorcee. Steiner has convinced her that in hot weather it makes sense to wear shorts, but typically she still prefers long pants over skirts or capris and lace-up shoes over sandals for better AFO camouflage.

"I can verbalize about the AFOs, but showing them off is another thing," she said.

In addition to her lower extremity issues, Covington also has diminished sensation in her fingers, but that hasn't stopped her from typing, quilting, and creating glass mosaics.

"I'm not at the point where I feel deprived because I have CMT," she said. "I'm just grateful that I'm almost sixty and it really hasn't impacted my life too much."





finding the right AFO makes a huge difference in their activity level and quality of life.

## Brennan Ryan: With AFOs and cool sneakers, this 10-year-old can be a regular kid

BY ROBYN PARETS

Photos by Joshua Albanese (joshuaalbanese.com)



If you ask Brennan Ryan what his favorite after-school activities are, he'll tell you, "Basketball, baseball, flag football, and Cub scouts." Ask him what his favorite part of school is and he'll answer, "Recess." This isn't surprising considering that Brennan Ryan is 10 years old and in fourth grade.



"He's a regular boy. He likes sports and has a ton of friends," said Kathy Ryan, Brennan's mother.

But, at the same time, Brennan, who has Charcot-Marie-Tooth disease (CMT, specifically subtype CMT1A), is no ordinary kid. His mother, who has extremely mild CMT symptoms, knew something was wrong from the time her son was six months old. He had trouble moving around and didn't start walking until he was around two years old. The developmentally delayed baby started physical therapy before his first birthday and, as a toddler, he had trouble with balance and couldn't run or

jump like the other kids, Kathy said.

Once diagnosed with CMT through genetic testing, Brennan immediately began wearing SureStep SMOs (supramalleolar orthoses) to help his excessive pronation and other symptoms.

"The SMOs helped his overall stability and gait. They also gave him confidence," Kathy said.

Orthotic devices have played a bigger role in Brennan's life as his CMT symptoms have become more severe over the years, said Kathy, who also has twin 12-year-old daughters who do not have CMT. Brennan becomes fatigued easily, frequently has pain in his feet and hands, and has trouble with weakness in his extremities. For example, his thumb dislocates often and he has difficulty holding a pencil. In school he uses technology, including an iPad with a keyboard, to help him write and keep up in class, said Kathy, who is a third and fourth grade teacher. Perhaps the hardest thing is watching her son come home from school wiped out or just lie on the floor after playing sports.

"It takes him a while to gain his energy back," she said,

Brennan goes to physical therapy once a week, and his mom also helps him stretch his legs at home. They've found that it's important for him to do these exercises regularly, Kathy said.

"He's a very bright boy and very aware of what he has [to deal with]," she said.

However, Brennan is happy to have orthotic devices that allow him to wear cool sneakers, just like his friends. Kathy and Brennan's father, Kevin Ryan, just have to search a little harder for the extra-wide foot-

wear brands that will fit over the devices.

"He wants the two-hundred dollar gym shoes that the other kids have," Kathy joked.

Fitting in socially is certainly as important as shoes that fit, at least if you're a kid from suburban Chicago like Brennan. At the same time, the Ryan family believes education about CMT and other neuromuscular disorders helps children accept differences among their peers. To this end, Brennan was happy to be chosen last year as a Muscular Dystrophy Association (MDA) Goodwill Ambassador for the state of Illinois.

"It was pretty cool. I got to do a photo shoot with a [Chicago] Bears player," Brennan said.

Kathy explained that her usually more reserved son came out of his shell to appear on MDA posters and marketing materials. He also made some public appearances. One of these included going to an elementary school to talk to the kids about CMT and other neuromuscular diseases.

"I liked it, and I think I helped other kids," Brennan said.

ROBYN PARETS IS A FREELANCE WRITER IN THE BOSTON AREA.



## *James Robinson: AFOs give him the confidence to stay active in retirement*

BY ROBYN PARETS

Photos by Susan Ruediger



Some may think a midlife diagnosis of Charcot-Marie-Tooth (CMT) disease might necessitate a sedentary retirement. Not James Robinson, who learned he had CMT1A when he was about 52 years old.



"I don't let it get me down or stop me from doing what I want to do," said Robinson, now a 75-year-old retiree who lives in the Atlanta suburb of Stockbridge, GA, with his wife Julia.

The consummate optimist is no stranger to adversity. In fact, he learned to deal with challenges long before he was faced with his CMT diagnosis. Growing up in an impoverished neighborhood of Pittsburgh, PA, Robinson never graduated from high school and instead enlisted in the US Air Force at age 17. His father was ill at the time, and Robinson felt that by joining the

military, he could learn new skills, serve his country, and help support his family. As it turned out, all of this proved to be true. Two months after leaving home, his father passed away. His mother was left alone with his three younger siblings. Robinson took over the role of family caretaker, even from afar.

He served in the Air Force for 24 years and became a computer systems manager. When he left the military in 1980, he settled in the Atlanta area. He and his wife raised three children and now have 12 grandchildren. Robinson worked for Georgia-Pacific as a computer systems specialist and kept in shape by jogging and playing racquetball.

Soon after Robinson turned 50, he began noticing that he was losing his balance often. His hands and legs were much weaker than usual. When he played racquetball, he couldn't control his swing, and he noticed that his gait was changing. Then he started to trip frequently, and fall down.

Robinson didn't understand exactly what was going on, but he knew something was wrong. Then he remembered that one of his nieces in Philadelphia had had similar symptoms since she was young. He contacted her, learned about her CMT diagnosis, and upon her advice, went to see a neurologist.

That neurologist sent Robinson home with no answers or diagnosis, so he sought out a second opinion.

"The second doctor ordered a test to detect nerve speed and diagnosed me. He sent me to a specialist at Emory [University], who confirmed it," he said.

Robinson's form of CMT1A is the most common of the more than 80 known sub-

types of the genetic neuropathy. He experiences all the symptoms typically associated with the inherited disease, including muscle weakness in the lower extremities and hands, high arches, numbness in the feet, and lack of balance. He has lost sensation in the tips of all of his fingers except his pinky fingers.

Although Robinson regularly does physical therapy to help alleviate some of his CMT symptoms, he said his Allard Blue Rocker ankle foot orthoses (AFOs) have made the biggest difference.

"My confidence level has increased significantly. I have more stability and energy. I would not be able to walk from one side of the house to the other without them. Until they find a cure for CMT, these are a necessity [for me]," Robinson said.

Robinson, who also suffers from chronic back pain caused by spinal stenosis, said that although he may not be able to swing a racquet anymore, he still can still fix things and do projects around the house.

"With my lack of dexterity, it takes me three times as long, and I have to use both hands to pick things up. I have to be more patient and inventive at times," he said. "But my attitude on life is to be positive."

ROBYN PARETS IS A FREELANCE WRITER IN THE BOSTON AREA.





*finding the right AFO makes a huge difference in their activity level and quality of life.*

**Jeff Seitzer:**

## *AFOs and staying fit help manage his mild symptoms*

BY ROBYN PARETS

Photos by Joshua Albanese (joshuaalbanese.com)



**It's hard to imagine that a 56-year-old man who swims, plays golf, works out, and even practices yoga has Charcot-Marie-Tooth disease (CMT). But Jeff Seitzer, who was diagnosed more than 40 years ago, has managed to keep his symptoms at bay. He believes staying fit and flexible has been the key to maintaining his active lifestyle.**



In fact, Seitzer says he noticed an increase in his stamina and energy level when he started attending yoga classes about 10 years ago. At home, he also stretches “maniacally,” and has noticed that his legs stiffen up when he doesn’t both stretch and exercise regularly.

Seitzer was about 13 years old when he was diagnosed with CMTX, which is typically associated with more severe symptoms in men than women due to its linkage to the X chromosome. Clinically, CMTX is similar to the more prevalent CMT1 subtype; common symptoms include muscle weakness and atrophy, and loss of feeling in the feet, lower legs, and hands.

Seitzer only has mild symptoms, which include foot drop, weakness, and pain in his legs and feet at times. Even his doctors at the CMT Clinic at the University of Iowa in Iowa City can’t believe how much he can still do.

“By my age, I should be hobbled,” Seitzer said.

Born in Omaha, NE, Seitzer now lives in Chicago with his wife, Janet Smith, and 8-year-old daughter, Penelope (who is adopted and therefore not at risk of inheriting CMT from Seitzer). The University of Chicago alum teaches religion and philosophy at Roosevelt University in downtown Chicago and often rides his bicycle to work. To help ease pain and extend his stride when walking, Seitzer has been wearing Kinetic Research’s Noodle AFOs (ankle foot orthoses) since 2014. Before that, he tried other types of orthotic devices, but he says the Noodle AFOs are a major step up for him. For example, they are particularly useful when wearing winter boots to walk on ice and in snow, as they help to improve his stability.

“They are a game changer,” he said.

Seitzer also said the Noodle offers support during other activities, like riding his bike. For instance, he said, when wearing the AFOs he can ride his bike without needing to stretch first. Also, the Noodle’s carbon fiber foot plate almost acts as a “biking shoe,” helping distribute his weight more evenly on the pedals.

If Seitzer had his Noodle AFOs 40 years ago, they might have saved him some embarrassment, too. That’s because, as a teenager, he wore metal braces. At the time, Seitzer’s mother, who also had CMT,

thought she was doing the best thing for him. She didn’t want him to end up like her father, who had a severe case of the disorder. As a result, Seitzer wore the awkward braces until he was about 20.

“They were not so great for those teenage years,” he recalled.

Seitzer’s mother was also hyper-vigilant about curbing the progression of her son’s CMT for another reason: At the age of four, he suffered from a major bout of encephalitis, which left him temporarily paralyzed on his right side. Other residual side effects left Seitzer with extreme nervous energy, memory loss, and mood swings. He also had reconstructive surgery on his right foot, which was malaligned and weakened by the encephalitis.

“It was devastating,” he said.

Despite a difficult childhood and his share of struggles, Seitzer considers himself lucky to be leading an active adult life.

“I have good and bad days,” he said. “I understand my body, and it’s an ongoing process.”

ROBYN PARETS IS A FREELANCE WRITER IN THE BOSTON AREA.



# Get Back Up Today!

### WHAT IS THE TREATMENT FOR FOOT DROP?

The most common treatment is an ankle-foot orthosis (AFO) which provides support to the ankle and foot. The AFO helps control foot drop and ankle instability by providing a better sense of balance. Often times, individuals are fit with a custom molded plastic AFO, however, there are other orthotic options available that provide superior function and performance.

The goal of orthotic treatment options is to help you maximize your mobility and independence. Chances are your most important need is to be able to walk better, without assistance and for longer periods of time without getting exhausted. Hundreds of thousands of people with foot drop have experienced a mobility rebirth thanks to the stability and dynamic assistance provided when wearing ToeOFF®, a unique patented carbon fiber composite AFO.

### WHAT IS TOEOFF AND HOW DOES IT WORK?

The ToeOFF product line is made of ultra-light weight materials including carbon fiber, fiberglass and Kevlar®. The light weight is especially important to those individuals affected with neuro-muscular deficits from CMT. ToeOFF provides a natural biomechanical response similar to the movement of your own muscles. The footplate and “open heel” design are major contributors to the function of the ToeOFF. When your heel strikes the ground, energy passes down the side of the AFO to create a dynamic response that reflects the energy to the footplate to prevent “foot slap” and lift up the forefoot, much like the spring of a swimming pool diving board. The design and materials allow for enough strength to control the position of your foot as you swing your leg, making walking easier with less energy consumption. Unlike a custom molded plastic AFO that covers and immobilizes the ankle, the ToeOFFs open heel design allows your heel to move freely as it normally would, allowing proper biomechanics to occur in the foot.

### WHAT ARE THE BENEFITS OF TOEOFF?

ToeOFF provides a stable, fluid, propulsive and symmetrical walking pattern. ToeOFF can improve your quality of life by:

**Restoring balance and improving stamina** – ToeOFFs strong and durable design allows you to move on uneven surfaces and climb stairs or ramps more confidently and independently, without stumbling or falling. Studies have shown that ToeOFF users can walk further in ToeOFF than in conventional molded plastic AFOs.

**Decreasing risk of muscle atrophy** - A recent study revealed that plastic AFOs can lead to ankle immobilization which resulted in calf muscle atrophy.

**Fits well into good support shoes without having to increase shoe size** – The thin and lightweight carbon fiber design ensures that you can wear your AFO inside any standard shoe that provides good support – without increasing your shoe size!

**Providing function with fashion** – Ask your orthotist about our Fantasy line that offers ToeOFF in ivory, purple, black or dark blue, or the removable SoftSHELL covers that are offered in beige or dark brown.

### HOW DO I OBTAIN A TOEOFF?

ToeOFF Products require a prescription from your physician and must be fit by a certified orthotist who will go through an eight-step customization process. The ToeOFF product line offers a variety of AFOs with graded stability to accommodate for different individual’s needs, sizes, and stabilizing properties. While the ToeOFF products are state-of-the-art in both materials and design, not every individual may be a candidate for ToeOFF. Allard USA offers a 30 day “Try It - You’ll Like It” Patient Satisfaction Guarantee for you to “test” that the ToeOFF will work for you. Talk to your physician about specifying “Allard ToeOFF - no substitutions” on the prescription and specifically ask for Allard ToeOFF with your Orthotist - this will ensure that you are offered the unique patented design of the ToeOFF. ToeOFF products are covered by Medicare and most insurance companies.

### YOUR FIRST STEP TOWARDS INDEPENDENCE

Finding the most appropriate AFO can be challenging so we encourage you to discuss your orthotic treatment goals and options with your physician, physical therapist, and orthotist. Taking an active role in your treatment plan will ensure that you receive the most beneficial and appropriate AFO that addresses your specific needs. There are many AFO choices that you will encounter - knowing your options and understanding the differences will ensure that you achieve the maximum success from your orthotic solution.



Virginia Mamone  
Diagnosed with CMT

[www.getbackUPtoday.com](http://www.getbackUPtoday.com)

Connect with others who have been or are currently experiencing similar struggles and successes as you. Become part of a supportive, encouraging group geared towards helping each other reach goals previously thought impossible.  
**Join the Movement and Get Back UP Today!**

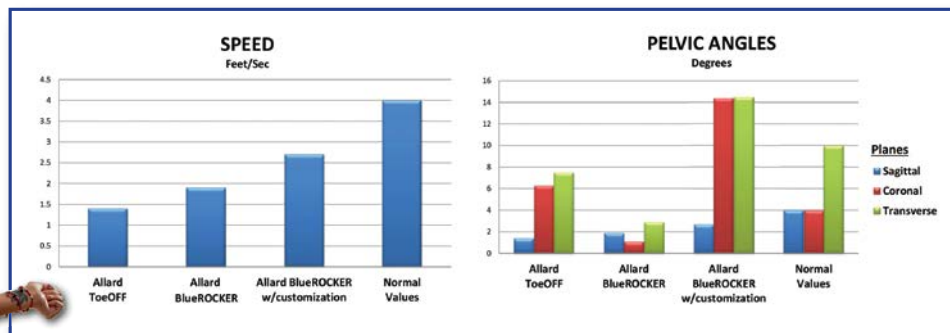
### FOR ADDITIONAL INFORMATION

Email [CMTApartner@allardusa.com](mailto:CMTApartner@allardusa.com)  
Call 888-678-6548 or visit [www.allardusa.com](http://www.allardusa.com)



# Case Study: Maximizing Functional Outcomes Utilizing Objective Gait Analysis

**Vincent DeCataldo, BOCPO, NJ LPO**  
 Manager Allard O&P Partnership, Allard USA



Allard ToeOFF Fantasy

Allard ToeOFF

## INTRODUCTION

Identifying maximal functional outcomes is often limited to visual gait analysis and subjective patient commentary, making it difficult to know if an adjustment or change of design has made a difference to function. Utilizing a portable gait analysis system, we are able to use quantified measures of gait to modify our components, designs, and gait training to maximize functional outcomes.

## PATIENT PROFILE

Subject is a 39 year old female with CMT and prominent symptoms of bilateral paresthesia and weakness in hands and feet. Patient was initially fit with bilateral custom posterior leaf spring (plastic) AFOs approximately 7 years ago. She rejected the plastic AFOs because they caused sores on her feet and she felt that it took more effort to walk with the orthoses than it did without. One year ago, she started wearing the Allard ToeOFF® and felt that they assisted her with activities involving long distances. She also tried the Allard BlueROCKER® and Allard Ypsilon™. Currently she walks, runs, trail hikes, and practices yoga on a regular basis and changes her Allard AFOs to give her more or less support and propulsion based on her activity.



## METHODS

Subject was tested in three different bilateral conditions. The custom fit dynamic carbon composite AFO designs included: 1) without customization, 2) with increased rigidity without customization, and 3) same rigidity as condition 2 but with customization to accommodate the heel height of the shoe. Temporal-spatial and pelvic motion data was collected utilizing a BTS G-Walk Portable Gait Analysis system.

## RESULTS AND DISCUSSION

Utilizing the G-Walk system, functional outcomes due to changes in orthotic design and customization can be measured and documented. The orthotic intervention that provided the maximum function for this patient happened to be the more rigid design (Allard BlueROCKER) customized for shoe heel height which allowed for increased speed and decreased pelvic motion. Speed and percent of double limb support were closest to normal values for women.

**Acknowledgements:** Thank you to Virginia Mamone and Brittany Stryker, OTD, OTR/L, BOCO and Orthopedic Motion

## TESTIMONIALS

*"I put them [Allard ToeOFF] on and my life changed. I no longer have to look at the ground when I walk... My Allard ToeOFFs have given my life back and are so liberating... Now I can go about my day very proudly with my head held high and with a whole lot of confidence."*

- Susan Ruediger, CMTA Director of Development, foot drop as a result of CMT.

*"I was unable to run and play with my children because of my drop foot. I was always tripping or could only walk a certain distance before getting tired. For a while, I wore plastic braces, but only for about a month and a half because they actually caused me more pain and fatigue than not wearing anything at all. In 2012, I found out about Allard through the CMTA and I got my Allard braces. And they have forever changed my life. I can finally do things with my kids like go to Disneyland, the grocery store, and shopping at the mall. Little things that mean the world to me. I am committed to being active and encourage others with either CMT or foot drop to do the same."*

- Virginia Mamone, foot drop as a result of CMT.

## REFERENCES TO PROVIDE TO YOUR PHYSICIAN, PHYSICAL THERAPIST AND ORTHOTIST

<sup>1</sup> Ramdharry, G., Pollard, A., Marsden, J., Reilly, M., Comparing Gait Performance of People with Charcot-Marie-Tooth Disease Who Do and Do Not Wear Ankle Foot Orthoses. MRL Center for Physiotherapy Research International, 17(4), pp 191-199. 2012.

<sup>2</sup> Dufek, J., Neumann, E., Hawkins, C., and Otoole, B., Charcot-Marie-Tooth AFO Mechanics and Gait Patterns. Lower Extremity Review. 2014.

# Carbon Fiber Orthopedic Braces

### AFO MECHANICS

Composite material advances over the past 10 years have improved the design creativity and possibility for clinical orthotists worldwide. In the past, an ankle orthosis required a completely solid and fully encompassing design in order to attain the mechanical control, durability and ultimately the function needed for corrective ankle varus bracing. These devices were essentially plastic shells that created a solid/static environment which was safer than not using a brace at all, but they were also highly problematic with patients complaining of pressure points and a clunky gait. Today we can simulate more natural gait patterns while completely avoiding pressure points.

### CONTROLLING THE TRI-PLANAR DEFORMITIES

Kinetic Research offers a variety of ankle braces, each with its own character and effect to meet the needs of the user, from the most basic needs to the most complex. For folks that have foot/ankle weakness but have good alignment of their ankle, we offer the Noodle line of AFOs. These are the most dynamic and least restrictive designs for controlling drop foot. When the user has a tight heel cord with mild supination or ankle varus, they become a candidate for a PLS design. When the user has more significant ankle varus he/she becomes a candidate for the ValgaNoodle AFO. The ValgaNoodle offers the highest degree of lateral control that we can provide. It's important to have a team approach when choosing the type of AFO that will work best for your individual needs, and there is always a trade off between control and mobility. The team is made up of the patient, the orthotist and the physician/therapist, and all have to be involved for the best results. The objective is to "put on as little as possible, but get the job done".

### IMPROVING STABILITY DURING GAIT

The ValgaNoodle is made specifically for varus control. It's a highly custom AFO, meaning that it is not assembled using pre-made components. The creation of a ValgaNoodle begins with a highly skilled orthotist who will evaluate and cast for the device. That attending orthotist will manipulate and control the patient's foot while he is casting, keeping the ankle in the best position possible to maximize the effect of the finished ValgaNoodle. Evaluations and observations made by the clinical orthotist are then incorporated into a design formulation that will work best for the individual patient. The design details are then determined based on the level of control vs free movement that is needed to maximize the performance of the user. One of the objectives is to keep the ValgaNoodle lightweight and as simple to use as possible. The orthotist will select the device based on the severity of the case, as well as the user's lifestyle,



Custom ValgaNoodle AFO with Pre-Tibial Shell - Model: NPV

activities and footwear.

### FITTING THE DEVICE

The one common mechanical concept for the ValgaNoodle variations of AFOs is, that they will all have a single medial strut with a floating lateral "reverse T" cuff.

#### Design options for the footplate:

1) Flat, in which we match the bottom of the shoe. When we make it in this format, it will usually have a custom foam removable arch support. It offers the least control but because it's flat, we can make it flexible.

2) Contoured. This is similar to a UCBL footplate. Our technicians will adjust the cast taken by your orthotist to increase lateral control in a compact way for easy shoe selection.

3) Contoured with high control. This system goes higher than the UCBL, encompassing the dorsum of the foot, and has a special 3-point strapping system to provide maximum control.

#### Design options for the strut:

1) Solid ankle, the highest level of control, removing the user's ability to dorsi/plantar flex.

2) Dynamic, which allows dorsi/plantar flexion, but with reduced lateral control.

#### Design options for the cuff:

1) Posterior, for a normal or hyperextended knee.

2) Anterior, generally used for knee extension weakness. Easier to don, but limited to low top shoes.

## Case Study:

**David B. Misener, BSc, CPO, MBA**

Clinician: Clinical Prosthetics & Orthotics

### INTRODUCTION

Many times I meet individuals with CMT who have had a long history of bracing in the past. They typically come with many positives and negative reviews of each and every device, and spending time reviewing these devices will help establish a successful plan for the new device. Completing a thorough evaluation will also include assessment of muscle strength, range of motion, balance and coordination, walking speeds, calluses, footwear, activities of daily living, goals, wants/needs, assistive devices used, etc. Typically gathering any kind of subjective and objective information may help in your decision-making process. The following is a case study describing the outcome of a Kinetic Research ankle foot orthosis.

### PATIENT PROFILE

Patient is a 59-year-old male with the diagnosis of Charcot Marie Tooth 1a. He comes from a family with a long history of extremely prevalent CMT. He is aware of his parent, siblings, children, nieces and nephews with CMT. He began wearing bilateral simple plastic posterior leaf spring AFOs in his mid 40s and then moved to off-the-shelf Allard AFOs in his early 50s. He's aware of the strengths and weaknesses of his body and doesn't let the CMT interfere with



his activities of daily living. He has had multiple Allard Toe-off AFOs which seemed to break after 4 to 6 months of his activities. He then progressed to the stronger Blue Rocker AFOs because of early breakage of the Toe-off AFOs and because he maintained a fairly high activity level. He was able to extend the life of the Blue Rocker AFOs but these also fractured in the 12-16 month time frame. His range of motion is within normal limits at all joints. He is able to dorsiflex to neutral but cannot dorsiflex beyond neutral at a strength of grade 2. His plantar flexors are a grade 3. He does not use any other assistive device for ambulation. As previously stated he is busy doing activities of daily living and often walks miles at a time along with hiking on the weekends.

He requests a smaller lower profile design along with some of the biomechanical advantages of the Allard systems. He is also hopeful that he can have a device that may be more durable and continue to facilitate his activities of daily living.

### AFO DESIGN

Given his overall successful history with off-the-shelf carbon AFOs it was decided that custom carbon AFOs that would maintain dynamic motion and energy return would be an excellent approach to offer him support while not restricting any portion of his activities of daily living. Together we selected the custom Kinetic Research Noodle anterior panel design. Kinetic Research also has many designs to choose from. In this case the approach was to maintain the anterior panel that he was proprioceptively used to and also to custom build a stronger material layup to reduce the likelihood of early failure. Once again there would be a soft removable foot orthosis to neutralize the pathomechanics of his foot. There would only be one strap across the posterior aspect of his calf to secure the device to his limb. He would rely on good footwear to stabilize the device around his foot and ankle.

### RESULTS AND DISCUSSION

When he first observed the devices he was pleased to see a smaller, lower profile design as compared to his traditional systems. Often this aspect of bracing design isn't emphasized as much as it could be but for the end user it is often very significant. He was pleased to see the smaller anterior panel and was also quite hopeful it would have the dynamic and energy storing properties to which he had become accustomed. While standing he was able to flex the systems significantly yet maintain control, balance and stability. With most CMT individuals I have observed it takes many steps and, for some, weeks before they can truly give accurate feedback on a new device. While ambulating he could feel the energy return in the devices and felt that these devices offered as much as he was used to. The custom removable foot orthoses were adjusted with mild lateral posting to fully stabilize his foot from heel strike to toe-off.

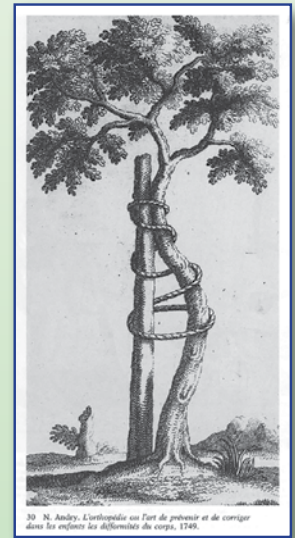
One of the main goals was to extend the life of a device, something that could only be assessed with the passage of time. At his initial fitting the devices seemed to be successful and he will return for multiple follow-up sessions within the next few months. At our first follow up session he reported that he transitioned into the new Kinetic Research AFOs without problems. He felt that energy return is on par with his previous systems and the smaller anterior panel felt more open and comfortable. He continued with his high functioning level of activities of daily living. At his one year follow-up there were only simple adjustments that were completed to return the device to its original design. This device still has held strong after three years of use and has allowed the recipient to continue with his goals and activities of daily living.



ValgaNoodle AFO



Custom ValgaNoodle AFO with Rigid Anterior Tibial Section



The Andry Tree Illustration

### AFO MECHANICS

The mechanics of the Valganoodle are classic for orthopedics in general. The Andry Tree shown above was an illustration from 1741 French publication *Orthopedie*. Just as it was illustrated 270 years ago, we still follow the mechanical concept of pulling the subject towards the stabilizing strut.

In the Valganoodle design, a floating lateral "Reverse T" cuff pulls the ankle towards its solid medial strut. We then use leverage and pressure in the calf and foot to create a 3-point pressure system that is very effective in controlling high level ankle varus conditions. The contoured footplate can be used to apply an additional 3-point pressure system to control the high arch / varus foot.

WADE BADER, KINETIC RESEARCH

## Specializing in advanced composite applications for the O&P industry

### AFO MECHANICS

The mechanics behind our family of Charcot Marie-Tooth (CMT) braces centers around the medial strut design. The medial strut folds into the footplate in the arch area, making the brace very stable when patients are in midstance. This provides the CMT patient with good standing stability. It also provides proprioceptive feedback higher up the leg to aid in controlled standing. We offer three proximal options to the medial strut design: anterior shell, which produces a ground floor reaction that stabilizes the knee; posterior calf, which aids in knee regravatum control; and our standard “T” top that aids in knee control in both directions.

### CONTROLLING THE TRI-PLANAR DEFORMITIES

Tri-planer control is achieved in the coronal plane with the use of a proximal strap and a distal supramalleolar strap. Further control can be achieved by using our CMT footbed, which has a lateral wedge running from the heel to the base of the fifth metatarsal head. For more control, a lateral half supramalleolar orthosis (SMO) can be used to provide maximum varus control by linking the SMO to the supramalleolar strap. This secures the lower extremity, creating a triangle of forces to control the CMT patient’s tendency to roll into varus. This is what gives our CMT patients good standing stability.

In the sagittal plane, midstance standing stability is achieved as the medial strut joins the foot plate. The strut is constructed with internal core material that makes it rigid without adding weight. The heel section of the AFO bends and deflects during walking to provide soft initial contact with the floor. This transitions to midstance. At toe-off the front of the footplate deflects, storing energy, then springs forward at the end of toe-off. The end result is an AFO that provides great midstance control without interrupting the rockers of gait. Proximal transverse plane control comes from the intimate fit that a custom-made brace provides around the tibia. Distally, our CMT footbed and SMO options provide rotational control while keeping the bulk of the AFO to a minimum in the shoe.

### IMPROVING STABILITY DURING GAIT

The initial benefit our AFOs provide is solving the foot-drop problem. This reduces energy consumption, as well as restoring the normal rockers of gait. This also improves balance. Because wearers don’t have to pick up each leg as high to clear the floor, their lateral center of gravity shift is reduced. Proprioception, or spatial awareness, is also improved because ground forces are transmitted to the top of the AFO, giving the patient accurate feedback about where their lower limbs are in space. This may not be initially appreciated but, after wearing our AFOs for a while, patients usually comment that they “can’t go without them” and “feel very unstable without their AFOs.”

The end result is that CMT patients can walk farther with less energy, walk normally (heel-to-toe gait), and have greater stability and confidence in any task they set out to do. Standing stability is also improved. Improved standing stability also reduces fatigue and allows the AFO wearer more freedom to just stand with a drink in their hand without looking for something to lean against. In summary, patients experience improved walking ability, improved proprioception and standing stability, reduced energy consumption, and greater confidence.

### FITTING THE DEVICE

Because all of our CMT AFOs are custom made, many of the optimizations are made during the manufacturing process. The information from the order form is used to customize each AFO. Options, such as the addition of ankle straps, alterations to the footbed, or SMO trimming, can be made by the orthotist. Most of the time the orthotist has little work to do when fitting our CMT AFOs.

### DEVICE WARRANTY

Our warranty against defects in materials and workmanship is for a period of six months from the date of manufacture. During the first six months a free replacement will be issued for the defective AFO upon its return and inspection of defect. One free replacement made from the original mold will be issued in a calendar year per patient. From seven to 12 months after manufacture, a prorated warranty will be used and charges will be adjusted accordingly. Skin contact parts (foam and straps) are warranted against defect in materials and workmanship for 30 days from the date of manufacture. Warranty against fitting issues, if accomplished within the first 30 days, are no charge. Our return policy is handled on a case-by-case basis. Another benefit of our AFOs is that minor problems can be repaired. This can be accomplished even after the warranty period for a reasonable fee and eliminates the need to replace the entire AFO.

## Case Study:

**David B. Misener, BSc, CPO, MBA**

*Clinician: Clinical Prosthetics & Orthotics*

### INTRODUCTION

Many times I meet individuals with CMT who have had a long history of bracing. They typically mention many positives and negatives of each and every device. Spending time reviewing these devices and the patient’s feedback will help establish a successful plan for a new device. A thorough evaluation also includes assessment of muscle strength, range of motion, balance and coordination, walking speed, calluses, footwear, activities of daily living, and patient goals and needs, as well as gathering information about assistive devices used. Gathering subjective and



objective information may help in your decision-making process. The following are case studies describing the outcome of a Custom Composite AFO and Kinetic Research AFO.

## PATIENT PROFILE

The patient is a 68-year-old man with the diagnosis of CMT1A. He is the first known case of CMT in his family. He has children in their 40s who may have CMT, but at this point the condition does not interfere with their activities of daily living and lifestyle. He has a 15-year history of wearing an AFO. Currently, he wears bilateral polypropylene semirigid AFOs that he has worn for approximately the last eight years. These devices are set in approximately 1° to 2° of dorsiflexion, with one proximal calf strap used to secure the devices to his limbs. There's no soft interface. He uses a single-tip cane to assist with balance and stability. He walks at a slower pace and is very focused on his balance and stability. He has significant muscle wasting distal to his knees; he is unable to actively dorsiflex (grade 0) his foot, and his plantar flexors (grade 1) are extremely weak. He has 1° to 2° of passive dorsiflexion range of motion around his ankle joint in subtalar neutral. His hindfoot can be brought to neutral, but there is no eversion of the calcaneus. He has mild plantar flexion in his first ray that causes his forefoot to drive his hindfoot into a few degrees of inversion. He has a subtle cavus foot with some callousing under the base of the fifth metatarsal.

He feels that his braces are too heavy and he would like to improve his balance, stability, and confidence, and also increase his activities of daily living.

## AFO DESIGN

After his long history of a semirigid AFO and the progression of his CMT, it was decided that a custom-carbon and equally supportive device would offer support to his weakened muscles and allow him to accomplish his activities of daily living with greater stability. The Custom Composite AFO was selected due to its light weight and strong design. Although Custom Composite has many designs to choose from, a stronger semirigid design was selected to achieve his goals. A removable insert was created to allow adjustability during the initial fitting and to fulfill future needs. A proximal calf and ankle strap was used to secure the device to the patient. Some of the subtle features of this design can have a powerful impact on the user with CMT. The light weight of the carbon AFO allows a smaller device with the same support as a heavier device. Although the energy expenditure is hard to calculate, it is often the first noted feature from the patient. The removable soft insert has lateral posting to reduce inversion instability and serves to translate the forces toward the first metatarsal. The addition of a strap across the dorsum of the foot allows the patient greater connection to the overall device.

## RESULTS AND DISCUSSION

When first standing the patient reported greater stability with the devices. He reported that this is largely attributable to the soft insert within the device because he feels his foot is more flat on the ground. The lateral postings serve to reduce the torque caused from his plantar-flexed first ray. Once his balance and stability was gained in a static position he began to walk. His initial comment was how much lighter the devices felt compared with his older polypropylene design. This always seems to be a very powerful feeling for the patient. He reported greater stability, confidence, and speed while ambulating. He also reported that he feels he is less focused on thinking about steps and believes he needs less support from his single-tip cane. Once final adjustments were completed he took the device home and will return in a few weeks for an evaluation of the new devices.

Patient currently reports he does feel that he is able to complete more activities of daily living throughout the day. He believes this to be attributable to the lighter weight, better control, and stability created from the design. He reports that he concentrates less and also makes fewer corrective steps as he walks. Clinical observation shows he walks faster and is using his cane minimally. His standing stability has greatly improved and he is much more confident ambulating. He will be followed up in six months.



**Hybrid medial strut with posterior calf**  
for patients who need knee control  
preventing recavatum



**Hybrid medial strut with  
anterior shell**  
for patients who need floor  
reaction to control knee  
instability



**Hybrid medial strut  
standard calf band**  
for patients who need knee  
control in both direction

## WHY A MEDIAL STRUT DESIGN IS IDEAL

Most CMT patients present with neurological deficiency causing foot drop, but later progress to a loss of balance. This loss of balance due to a decrease in proprioception leads to a constant readjustment of foot position to maintain standing balance.

### A Revolutionary Bracing Concept

#### AFO MECHANICS

SureStep manufactures a variety of products but is primarily known for the SMO (Supra malleolar orthosis) and pediatric AFOs (ankle foot orthosis). The SureStep SMO is meant for patients up to 80 pounds with hypotonic presentations. The purpose is to control the side to side movements of the ankle. The trim lines can be extended longer laterally or medially depending on whether the patient is pronating or supinating. As patients grow older there is a device called the Big Shot that accommodate children and adults weighing over 80 pounds. What makes the SureStep SMO so unique is the dynamic function that the brace allows. It uses compression to create alignment, unique trim-lines to allow freedom of motion where needed, and made from a material that travels together along with the foot.



SureStep BigShot

#### CONTROLLING THE TRI-PLANAR DEFORMITIES

This works under compression to maintain stability through midline. In young patients with CMT they can possibly present with a more over pronated foot position because of the low muscle tone (hypotonia) prior to developing a high arch foot presentation. This can be seen in CMT patients who have some symptoms of the disease but have not developed a high arch otherwise known as pes cavus foot. The SMO holds the heel in vertical alignment, giving stability in a coronal plane (side to side) position. An SMO is not necessarily for a patient who has weaknesses pushing up or down the ankle, just poor side-to-side movement.

#### IMPROVING STABILITY DURING GAIT

The device is designed to allow for motion to still occur from in and out of the midline position but limits the extremes that the foot would go to, in other words the extreme rolling inward or outward of the ankle. The goal is to improve balance and stability, or limit orthopedic injury. This helps limit out toeing or in toeing as well by improving the position to foot is landing on the ground. Having improved alignment may also utilize remaining strength for dorsiflexion or plantarflexion because the line of pull that these muscles are in has improved line of progression.

#### FITTING THE DEVICE

The trim-lines of the SureStep SMO are designed to be either longer on the lateral border for pronation or medial border for patient who supinate excessively and the orthotist should make the appropriate selection based on the patient's mechanics. These trim-lines are critical to the successful outcome of this device. Although the SureStep SMO is primarily designed for a younger population as children get older or adults who still have need for such a device the same mechanism of function can be accomplished using the BigShot SMO manufactured by SureStep.

#### DEVICE WARRANTY

SureStep products are guaranteed for fit, materials, workmanship and growth for ninety (90). During that time, should there be a problem, we will adjust, repair or replace the brace at no charge.

#### Case Study:

Sean McKale - CO

Midwest Orthotics & Technology Center

#### INTRODUCTION

Children with developmental delays with familial history of Charcot Marie Tooth (CMT) disease should be looked at with careful considerations. Developing early gross movement patterns are important in maintaining functional movement throughout life. CMT can have associated lower muscle tone, and despite muscles still having strength to move the foot and ankle muscle tone may affect the resting posture that the person is aligning. Maintaining improved biomechanical alignment can help facilitate motion in the muscles as they continue to function, prevent orthopedic injury, limit fatigue, and increase stability. Depending on the level of impairment will determine the necessary level of orthotic prescribed.

#### PATIENT PROFILE - SURESTEP BIGSHOT SMOS

Patient is a 5-year-old male with the diagnosis of Charcot Marie Tooth 1a, whose 35-year-old father is also diagnosed with CMT and is now wearing AFOs. He is being evaluated for orthotics because he has trouble keeping up with other children his age. He has no orthotic history, but because of his father's experience the family felt orthotics could be beneficial. He was delayed in walking as he did not take his first steps until almost 2 years of age. Currently, he has difficulty jumping and running, which are his primary complaints. Due to low muscle tone, his ankle collapses into an overpronated foot position. He stands with a wider base of support, and his feet appear to be externally rotated below him. He has the ability to lift up his ankle into dorsiflexion, and step up onto his toes. However, he has poor endurance to do so. When he attempts to jump, his feet do not fully elevate off of the ground.

Patient's ankle position is correctable to neutral alignment with good flexibility. Primary orthotic goals will be to maintain neutral coronal foot alignment. An SMO was discussed with the family versus a foot orthotic, because of the advantage of lever arm that the orthotic has.

#### SMO DESIGN

Patient was fabricated bilateral SureStep Bigshot Lite SMOs. This brace offers correction through compression. It also allows for freedom of the forefoot to come into contact with the ground, which can help proprioception and intrinsic muscle positioning. By correcting the coronal foot position (side-to-side) this will allow



SureStep SMO



the patient to better utilize the muscles that are responsible for lifting up and pushing downward at the ankle. An example of this is, it is much easier to pull a garden hose straight on versus pulling it around the corner of your house. The Bigshot Lite was selected because of the patient's weight and activity level.

## RESULTS AND DISCUSSION

When the patient was first fit with SMOs, he remarked that they were comfortable to use, had improved heel-toe progression, and had improved ability to push up onto his toes in standing.

Patient was seen again at 6 month follow-up for strap replacement. Braces were still fitting appropriately, and would anticipate child at this age to wear SMOs for one year or slightly longer. Patient had improved ability to jump and run. Parents remarked that they noticed improved differences in his playing abilities and keeping up with peers. Plan was to continue wearing SMOs until he outgrew them to maintain quality of movement and develop further patterning of gait. Anticipation to progress through a foot orthotic as he outgrows them to maintain joint stability and improved biomechanics.

This patient may require use of AFOs as his CMT progresses. But having improved alignment will make him more manageable in the future and especially in early childhood developing movement patterns is particularly important. It is known that we develop our adult gait pattern by the age of 7, so early intervention can be very important in developing improved biomechanics.

*Male patient before and after correction with SureStep BigShot SMO's*



SureStep SMO



# Say Hello to Gretchen

Gretchen has CMT. She relies on the CMTA to find out as much as she can about CMT. If you know people like Gretchen, you can find out about the latest research and advances at [cmtausa.org](http://cmtausa.org).



WEST GEORGIA

