DIAGNOSTIC INSIGHTS

ANALYZING GAIT CHANGES

ORTHOTIC MANAGEMENT

CLINICAL CASE STUDIES

Lower Extremity Review

ler

In Step With

Pediatric Hypotonia

Sponsored by an educational grant from SureStep

LER SPECIAL EDUCATIONAL SERIES

Lower Extremity Review
ler

In Step With
Pediatric Hypotonia

Contents

FROM THE EDITOR:

Bridging the Gap

For a parent, the only thing more frustrating than knowing something isn't quite right with your young child is hearing that nothing can be done until the underlying cause of the symptoms is identified. Luckily, parents of children with hypotonia can be spared the latter frustration.

As we detail in this special publication, early intervention in children with hypotonia—sometimes as early as in the neonatal intensive care unit—is not only possible but strongly recommended by experts, even in the absence of an underlying diagnosis.

A combination of physical therapy and orthotic management can help toddlers with hypotonia overcome developmental delays in gross motor skills, sometimes to the point of surpassing the development of unaffected children. Clinicians have been seeing these types of positive outcomes in children with hypotonia for years, and now researchers are beginning to quantify and document the results.

But too many pediatric lower extremity practitioners lack a detailed understanding of hypotonia, its effect on gait, and the therapeutic options that are available. This special issue, packed with evidence-based information as well as personal success stories, will go a long way toward bridging that gap.

And that means more children with hypotonia, with or without underlying diagnoses, soon will be cruising, walking, running, and playing right along with their peers.

BY JORDANA BIEZE FOSTER, EDITOR

BY SUZI KLIMEK

An unexpected path, an invaluable perspective

FOR THIS FAMILY, ONE TINY EXTRA CHROMOSOME LED TO A JOURNEY OF SELF DISCOVERY

When our son was born, we prayed for a healthy baby with 10 fingers and 10 toes. Our prayers were answered. Three years later, those same prayers were said for baby number two. And, once again, our prayers were answered. Baby number two was also born with 10 fingers and 10 toes. But there was one microscopic addition: an extra copy of chromosome 21. Otherwise known as: Down syndrome.

Being told that your child was born this way—it was a shock. It's just not something you think will happen to you.

I remember when the doctor came into my room. He sat down next to me and explained that our daughter likely had trisomy 21. I asked, "What is trisomy 21?"

At the time, my husband was at home with our 3-year-old son. I called him crying and confused. When he arrived at the hospital, he reassured me that everything was going to be OK.

"Julia is our daughter, and we love her unconditionally, so let's just move forward and figure it out as we go," he said.

I can't say that I rolled with it quite the same way. I cried—a lot. I kept asking, "Why us?" People kept telling me, "God doesn't give you more than you can handle." And I would reply, "Well…I think God confused me with someone else!"

It wasn't until I received the first sympathy card that it all made sense to me. I tried not to be offended as I knew the intention was good. It was then that I realized I didn't have any reason to be sad. I was, and still am, blessed to have this child. She is our winning lottery ticket. But that didn't mean it would be easy.

Within days of Julia's birth, we were connected with the Indiana First Steps Early Intervention Program. Julia started physical therapy at four weeks old. Developmental, occupational, and speech therapies soon followed. Julia was not born with any additional medical needs. However, because most children with Down syndrome have hypotonia, or low muscle tone, we knew that needed to be addressed. Because of the hypotonia, our physical therapist said that Julia would benefit from supramalleolar orthoses (SMOs) to help her walk. She specifically recommended SureStep.

We are lucky to live near the SureStep corporate headquarters. Julia was 18 months old when she first started wearing SureStep SMOs, and she was walking shortly after that. Since then, she hasn't stopped. Now 9 years old, she loves to play soccer, ride her bike, and climb on the playground jungle gym.

It's funny how things happen for a reason. If it weren't for Julia, my path would have never crossed with Bernie Veldman, owner and developer of SureStep, and I wouldn't be the company's executive director of marketing. I'm thankful that I can work for a company that is so passionate about helping children with special needs.

As people in our community meet Julia, they are drawn to her charming personality. She is a celebrity of sorts. At a local clothing store that we frequent on mommy–daughter days, one of the employees adores Julia and always asks her to model her clothes when she tries them on. Our son's baseball team has adopted her as a member of their team.

It's no different at school. Her classmates high-five her in the hallway, help her with her backpack, and open the door if she needs assistance. They even write her letters to tell her that she is their best friend.

Julia is too young to understand her celebrity status, but I know that all the love and attention she receives gives her the confidence she needs to develop into an even more amazing young lady.

As I look back on these nine years, I wonder why I ever shed a tear about having a child with Down syndrome. It was the path our family was chosen to take, and I know we're up for it. I could not imagine life any other way. Without a doubt, our family and friends have been profoundly affected by this gift. The people we've met, the challenges we've faced, and even my career choice have all been influenced by one little girl.

Her name is Julia.

Suzi Klimek lives with her husband and two children in Granger, IN. Originally from Wayzata, MN, she relocated to Granger after graduation from Saint Mary's College in Notre Dame, IN. Klimek is the executive director of marketing for SureStep.

A version of this article was previously published in the June 2011 issue of *Family* magazine.

BY CHRISTINA HALL NETTLES

Understanding Hypotonia

Diagnostic challenges should not delay clinical intervention

Hypotonia, or abnormally low muscle tone, is by itself not a disorder but a symptom of an enormous array of issues—many of which can be difficult to diagnose accurately. Even in the absence of a specific underlying diagnosis, however, children with hypotonia can benefit from clinical intervention.

Hypotonia can result from damage to the brain, spinal cord, nerves, or muscles, or may be a result of genetic, muscular, or central nervous system (CNS) disorders. The condition appears independently from muscle weakness, although the two may coexist in some disorders, such as motor neuron disease or multiple sclerosis. Young children with hypotonia appear "floppy" and may present with inappropriate head lag, astasis, hypermobility, decreased deep tendon reflexes, and problems sucking or swallowing. Older children with hypotonia may exhibit delays in gross motor skills or coordination or problems with ligament and joint laxity, respiratory control, posture, or speech.[1,2]

Demographically, hypotonia affects both genders equally and is no more likely to occur in one particular ethnic or racial group than another. It is one of the most common muscular abnormalities diagnosed in newborns with disorders such as Down syndrome, cerebral palsy, Prader-Willi syndrome, and Tay-Sachs disease yet, in some connective tissue disorders or certain muscular dystrophies, low muscle tone may not be revealed until later in life. Hypotonia is an associated symptom among many

> *"Hypotonia can be a symptom of more than 600 genetic disorders, with still more waiting to be identified."*

children with autism spectrum disorders,[3,4] but typically improves over time and responds to clinical intervention.

Neuromuscular specialist for children Thomas O. Crawford, MD, who treats patients at Johns Hopkins Children's Center in Baltimore, MD, estimates more than 95% of patients he sees for hypotonia can be evaluated with a comprehensive history and physical examination.

"In central issues, muscle tone may be diminished, but there is a discrepancy between tone during power of motion and the resting tone. That's an important distinction. If I find a child has more vigorous muscle power than tone, that leads me to concentrate on CNS issues," Crawford explained.

Expertise matters in both performing diagnostic studies on children, such as EMG, and assessing individual abilities, he said.

"If a child is weak as well as hypotonic from a central cause, his face is less animated than, for example, a child with spinal muscular atrophy, but in children with congenital myopathies, their faces may look unresponsive but the children are not," Crawford said.

In the 1970s, neurologists began classifying types of hypotonia, but "benign congenital hypotonia" remained a diagnosis when no cause could be found. Improvements in identifying genetic, neuromuscular, and connective tissue disorders now provide more detailed answers, opening the door to appropriate treatment responses. Still, some families live with the frustration of knowing hypotonia exists without discovering its underlying cause.

In some cases, idiopathic hypotonia resolves within the first few years of childhood, though minor cognitive impairments or developmental delays may persist.[5] Hypotonia caused by hormonal or metabolic disorders, such as rickets or congenital hypothyroidism, must be specifically screened for but can be easily treated.

Ronald D. Cohn, MD, chief of the Division of Clinical and Metabolic Genetics at The Hospital for Sick Children in Toronto, Canada, and an internationally recognized specialist in the genetics and clinical care of children with hypotonia, helped design a diagnostic algorithm to streamline hypotonia assessment for clinicians, distinguishing primary involvement of the upper motoneuron (central hypotonia) versus the lower motoneuron and motor unit (peripheral hypotonia) to indicate, for example, the need for magnetic resonance imaging.

In their 2011 study,[6] Cohn and genetic counselor Emily C. Lisi, MS, concluded that hypotonia can be a symptom of more than 600 genetic disorders, with still more waiting to be identified.

"A staged diagnostic approach categorizing patients as having peripheral, central, or combined hypotonia is the most efficient to providing a rational work-up. Establishing a diagnosis is crucial for prognosis, management, and treatment strategies and for ascertaining an accurate recurrence risk for future offspring," they wrote.[6]

Hypotonia in chromosomal abnormalities

"Individuals with Down syndrome are at risk for foot alignment problems due to hypotonia and ligamentous laxity. Both characteristics contribute to joint hypermobility," said senior physical therapist Patricia C. Winders, PT, director of therapies at the Anna and John J. Sie Center for Down Syndrome at Children's Hospital Colorado in Aurora.

"The ligaments do not hold the bones together tightly for optimal alignment and function. The joints of the foot have excessive flexibility, which causes instability and inefficient mechanics when standing, walking, running, and jumping," she said. "The child cannot use his strength effectively because the muscles are not aligned for efficient activation. Since his strength does not generate efficient power, he uses more energy during each skill and fatigues more quickly. The consequences of faulty alignment and mechanics range from impaired performance to pain, which can result in limitations in walking. Since walking will be vital for his entire lifetime, it is very important to be proactive in promoting optimal alignment and function, beginning when he learns to walk."

The goal of physical therapy for an individual with Down syndrome is to achieve maximal physical potential and to build a body that is fit and functional throughout his or her life, Winders said.

"Because of physical problems [hypotonia, ligamentous laxity, and decreased strength], he is prone to develop compensations, which are ways he adapts to make up for the physical problems. Some compensations, if allowed to persist, will eventually result in inefficient and painful movement patterns that will compromise his function as an adult," she said. "Physical therapy needs to focus on minimizing

the compensations that will lead to impairment of motor functioning and on building the posture, strength, and movement patterns that he will need as an adolescent and adult."

Hypotonia is a common feature of genetic disorders associated with developmental delay. Children with Angelman syndrome have gait ataxia with tremulous limbs, but may not always have hypotonia.[9] Neonates with Prader-Willi syndrome (PWS), however, have severe hypotonia, evidenced by lethargy and weak or no sucking. As children grow, their gross motor skills are typically delayed; they may sit at 12 months and walk at 24 months, though in some cases walking may be delayed until they are aged 4 or 5 years.

The insatiable appetite that is the hallmark of PWS does not typically present before preschool age, but obesity rates begin to soar during preschool years. Scoliosis; hip dysplasia; respiratory control issues; early risks of osteoporosis; short stature; short, wide feet that require extra care in fitting shoes properly; and the stress of a restricted dietary and home environment pose challenges for the families of children with PWS and their healthcare providers.[10]

In 2010, Korean researchers found a high prevalence of spinal deformity, limb malalignment, and foot abnormality in PWS, regardless of age or obesity.[11] They urged pediatric orthopedic surgeons to evaluate PWS patients annually for these conditions because of their possible concealment by obesity.

Early intervention with occupational and physical therapy and lifelong strength training and aerobic conditioning are critical to addressing overall health status, yet therapists must be aware that many patients with PWS have decreased sensitivity to pain. Any evidence of pain should be promptly addressed, as it may suggest a serious but masked underlying problem, such as a fracture or abnormality.

Missed diagnoses

Cohn suggests children with mild hypotonia and features such as joint hypermobility, pectus excavatum, pes planus, or cardiac abnormalities be carefully evaluated, as he believes patients with connective tissue disorders who have less severe forms of hypotonia remain dramatically underdiagnosed.

Crawford, too, cautions that, beyond the parameters of classic presentations of obvious congenital disorders of infancy, hypotonia may be missed.

"Children with classic Duchenne muscular dystrophy are not hypotonic as infants and, as a consequence, that diagnosis may not be considered until later. Some pediatricians might not think of Duchenne dystrophy for a boy who at 15 months of age manifests motor and cognitive delay. But weakness doesn't show up until later. In many of those cases, we are missing diagnoses," Crawford said. "Any boy not walking by 16 months should have a CK [creatine kinase] screening. In the case of Duchenne, the result will be five digits even though the child may show no signs of weakness."

Congenital hypotonia may be seen in tandem with joint hypermobility or ligamentous laxity, but, excluding obvious dysmorphic presentations, hypermobility may not be diagnosed until children are school-aged and present with arthralgia, back pain, abnormal gait, or joint deformity. Knees, elbows, wrists, metacarpophalangeal joints, and ankles are most commonly involved, according to British arthritis researchers, who collected data during a three-year period from pediatric rheumatology and hypermobility clinics.[12] Nearly half of the study participants were described in their clinical history as "clumsy," and more than a third showed signs of poor coordination in early childhood.

Dutch pediatric physical therapists performing a retrospective study concluded that one-third of children with generalized joint hypermobility presented with severe delays in motor development, though there was no significant association between the number of hypermobile joints and the age of independent walking.[13]

Joint hypermobility, if associated with hypotonia, may be indicative of Marfan syndrome, Ehlers-Danlos syndrome, osteogenesis imperfecta, or other mild variants of these and other musculoskeletal or connective tissue disorders.

Objective assessments

In a 2005 study, physical therapists and occupational therapists found that the specific characteristics of hypotonia included increased flexion, hypermobile joints, round shoulder posture, decreased strength, low activity tolerance, delays in motor development, and poor attention and motivation.[14]

Some studies acknowledge that objective measurement of the degree of hypotonia in an individual poses a challenge because, historically, therapists have defined resistance subjectively based on their methods and areas of expertise. Reliable tools, such as the 2011 Segmental Assessment of Trunk Control (SATCo),[15] which

> *"An inability to define an underlying diagnosis for low muscle tone should not interfere with the ability to manage hypotonia in patients of any age."*

measures discrete levels of trunk control in children with motor disabilities, are being developed.

Billi Cusick, PT, MS, C/NDT, COF, of Telluride, CO, uses SATCo in her practice.

"In my experience managing the alignment and movement disorders that are common in children with hypotonia, the functioning alignment of the joints and the somatosensory system are critical areas of concern," Cusick told *LER*.

She pointed to a recent systematic review that determined children with benign joint hypermobility syndrome demonstrated significantly poorer proprioception compared to children without the disorder.[16]

"Mechanoreceptors in load-bearing joints and the skin on the plantar foot deliver sensory information about limb position and weight bearing when they are stimulated," Cusick explained. "When joint surfaces are malaligned due to laxity in supporting ligaments, those receptors lose appropriate contact, and, presumably, their messages to the central nervous system are compromised. Lax joints fall to end range when loaded, where it appears that the sensory receptors finally detect the functioning positions. This lack of adequate and timely sensory information is evident in postural deviations, such as a wide-based stance and gait, excessive spinal lordosis, anterior pelvic tilt, knee hyperextension, and foot pronation seen commonly in children with hypotonia. For these children, such postural deviations are normal, and they persist without intervention."

Intervention with physical therapy and orthoses in childhood is key to preventing or managing pain in adolescence and adulthood commonly associated with joint hypermobilility and hypotonia, even if an underlying diagnosis is not determined. Persistent postural deviations and foot joint laxity typically deform the feet and interfere with independent mobility and endurance as body size and weight increase, Cusick said, citing a 2011 study by Wolf et al that recommended managing the pain with prolonged therapy and general conditioning, with special emphasis on improving strength and proprioception.[17]

Cusick not only has decades of experience treating children clinically, she has used her expertise with her own daughter, Ting, who arrived from China aged 12 months and unable to assume an all-fours position, crawl, get into and out of a sitting position, or pull to kneeling or standing.

"Her hips were very weak," Cusick said. "She had never taken weight on her knees or feet before, so I began to help her put weight through them in small increments and in a variety of postures. I used a jumper seat suspended on a spring and later fitted her in a pair of support shorts to keep her hips from sliding into full abduction when she attempted to assume all fours."

Ting was soon crawling and pulling to stand, but her feet were profoundly pronated, her foot ligaments were lax, and her wide stance imposed further pronatory forces on her forefeet. Cusick fitted her with heel cups and sturdy flat-soled sneakers, and Ting was soon cruising. At age 5 years, she began gymnastics; at 6, she started soccer; and at 15, Ting is now a competitive cheerleader.

"Although Ting's feet are aligned and competent, because she has a tendency toward joint laxity that is typical of the Chinese population, she continues to wear plantar orthotic inserts that protect her feet and knees from the wear that commonly occurs with excessive pronatory strain," Cusick said.

In caring for patients with low muscle tone or ligament laxity, Cusick relies on management strategies that improve functioning joint alignment and raise the level of—and improve the quality of—sensory input in daily life. To support these goals, she developed TheraTogs, a live-in orthotic undergarment and strapping system for children with hypotonia and other issues that is designed to deliver enhanced sensory input and improve postural alignment, and can be used in conjunction with orthotic devices that specifically target the foot and ankle. She also selects play activities that build balancing skills, muscle strength, and muscle tone, while maintaining the feet, trunk, and hips in optimum alignment.

As Cohn and Lisi underscored, an inability to define an underlying diagnosis for low muscle tone should not interfere with the ability to manage hypotonia in patients of any age. Symptomatic treatment can and needs to be tailored, they wrote, to create lifelong strategies vital to maintaining strength, reducing pain, and fostering independence.

CHRISTINA HALL NETTLES IS A FREELANCE WRITER BASED IN MONROEVILLE, AL.

References are available at lowerextremityreview.com, or by scanning the QR or tag codes at left.

BY CARY GRONER

Gait: The Cornerstone of Intervention

Quantifying the effects of hypotonia starts in the clinic

Effective management of children with hypotonia requires an understanding of how the condition affects gait. Clinicians typically rely on their professional experience when discussing the effects of hypotonia on gait in pediatric patients, partly because they trust that experience, but also because so little research has actually elucidated these effects.

"This needs to be explored from a research standpoint, because we really don't know," said Mark Holowka, CPO, who practices at Children's Healthcare of Atlanta. "It's difficult to quantify hypotonia because muscle testing isn't reliable in such young patients. As a result, we tend to rely on what we can observe and what we hear from the parents and the physical therapists on the team."

Pending better funding for formal studies into hypotonia and associated hypermobility, many clinicians feel they should conduct informal research as best they can.

"I think there has to be a real emphasis placed on gait analysis within clinical practice," said Jason Henry, MSPT, LO, the practice manager at Hope Orthotics in Spring, TX. "We have to take the mindset that clinical practice is research. Many different aspects of gait can be analyzed even if you don't have a gait lab, and that data has to be collected on a broader scale. We need more information about outcomes."

According to Holowka, a grasp of the distinctions between pathologies associated with hypotonia is critical to clinical intervention, regardless of similarities in symptoms.

"For example, autism is more of a sensory issue, whereas in a Down syndrome patient, it's important to focus on their joints and joint laxity," he said. "You have to understand what is associated with each pathology, because these kids will have different developmental and physiologic concerns."

Holowka added that performance inside the lab can differ significantly from that outside—a phenomenon he's found more prevalent with autism patients than others. Parents are often the best source of insight about such disparities.

"The patient will walk perfectly in the lab, but then the parents say, 'Oh no, they don't walk like this at home,'" he said. "It's ironic, because ideally what we want is for them to walk imperfectly in the lab so we can help them walk perfectly outside of the lab."

Kathy Martin, PT, DHS, a professor and director of the Doctor of Physical Therapy program at the University of Indianapolis, agreed that clinicians must rely on their own observations and measurements to characterize gait in children with hypotonia. And, despite the paucity of formal studies, most clinicians seem to agree about the outlines of those characteristics.

"My opinion is that hypotonia affects gait, in that I see kids with a wider base of support and more cocontractions, who are trying to artificially create stability," she said. "These kids are using everything but the kitchen sink to keep themselves upright because they have to work so hard to control their body against gravity."

Such patients may be less able to adapt to environmental obstacles than typically developing children, Martin explained, and have to change their ambulatory strategies when confronted with such obstacles.

"Children with Down syndrome may drop to their knees and crawl over an obstacle rather than stepping over it," she said. "Down syndrome also includes issues related to balance and postural control, of course; it's not just hypotonia. That raises the issue of what is the limiting factor, why they feel they can't step over the object—but hypotonia contributes to that."

> "Down syndrome has become the research standard because hypotonia is more consistent in that patient group."

The reading

In the scant literature that does address hypotonia in pediatric patients, a few key points are worth noting. First, though hypotonia is often thought of as muscle weakness, researchers point out that it is more accurately described as reduced resistance to passive range of motion in joints. Weakness, by contrast, is defined as a reduction in the maximum muscle power that can be generated.[1]

Many neurological conditions have been associated with hypotonia; the most commonly studied connection is with Down syndrome (DS), but others include Prader-Willi syndrome (PWS) and Ehlers-Danlos syndrome (EDS). Although hypotonia does occur in children who do not have a specific neurological diagnosis, the difficulty of categorizing such children means they are less likely to be included in scientific studies.

According to Faye McNerney, PT, DPT, a pediatric physical therapist in Troy, OH, DS has become the research standard because hypotonia is more consistent in that patient group.

"There is no gold standard for describing hypotonia, and because everyone with Down syndrome has some degree of hypotonicity, they are chosen as research subjects," she said. "Children with low muscle tone typically have joint hypermobility, which results in various biomechanical effects. Hypermobility of the foot-ankle joint allows the feet to pronate, and with time you'll see shortening of the peroneal muscle, lengthening of the posterior tibialis, medial rotation of the tibia, genu valgum, and hip internal rotation. As a result of these issues and poor balance, patients typically use a wide base of support during stance and ambulation."

Curt Bertram, CO, National Orthotic Specialist for Hanger Orthopedic Group who works at Children's Hospital of Wisconsin, in Milwaukee, agreed.

"When I talk about hypotonia, I think about Down syndrome patients, because those are the quintessential hypotonic patients," he said. "They have a hard time with advanced ambulation skills—climbing stairs, negotiating ramps, and running."

Indeed, because DS has been studied more than other syndromes, a review of the literature can shed at least some light on hypotonia and gait. For example, early research in DS patients reported that characteristic gait deviations included increased variability, out-toeing, flat foot at initial contact, a wider base of support, and poor foot control.[2]

A more recent paper by Italian researchers, published in *Gait & Posture* in 2008, further clarified aspects of gait associated with DS. Compared with controls, 98 children with DS (mean age 11.7 years) walking at a self-selected speed had more hip flexion during the whole gait cycle, more knee flexion in stance phase, less knee range of motion (ROM), and more ankle plantar flexion at initial contact. Moreover, ankle power was limited in terminal stance and preswing, as indicated by low propulsive capacity at push-off. Children with DS also had stiffer hip joints but more lax ankle joints.[3]

In a 2012 study of foot-ground interaction during upright standing, some of the same researchers found that 99 children with DS (mean age 9.7 years) exhibited larger midfoot and reduced forefoot contact areas, increased arch index values, and increased average contact pressures in the midfoot and forefoot than controls.[4] Of course, standing measures don't necessarily predict gait mechanics, but the authors noted that the prevalence of flatfoot in DS patients, associated with higher contact pressures, suggested the importance of clinical observation and intervention to reduce balance and gait impairment. Some gait impairments, however, would appear to be less affected by foot posture.

For example, Brazilian researchers compared gait in toddlers with DS with controls and reported no differences in stiffness or lower limb cocontraction indices (CCIs) during stance; however, DS

patients showed greater CCIs during swing phase.[5]

In 2011, Italian researchers also evaluated gait patterns in Ehlers-Danlos and Prader-Willi syndromes. Although this research was conducted in adults, the rare conditions have been so infrequently studied that results are worth noting here.

The two conditions were associated with different gait strategies. Patients with PWS showed some functional limitations at every level of the lower limb joints, whereas those with EDS had greater limitations that were nevertheless found mainly at the distal joints.

The authors recommended that PWS patients be encouraged to walk to improve muscle mass, strength, and energy balance, while rehabilitation for EDS patients should focus on improvement of ankle strategy.[6] Another Italian study comparing gait patterns in adult PWS and DS patients reported a cautious abnormal gait in both groups that was even less stable in DS patients. Those with DS also demonstrated significantly less hip flexion, knee flexion, and ROM in all joints compared to PWS patients. Both groups had significantly weaker push-off than controls.[7]

Finally, Canadian researchers reported in 2011 that children with autism had significant differences in cadence and peak hip and ankle kinematics and kinetics compared with controls. The children with autism also had reduced plantar flexor moments and increased dorsiflexion angles, which may have been associated with hypotonia.[8]

As noted, clinicians have long observed similar patterns in their patients with hypotonia.

"The disruption in neurology between the feet and the brain leads to an inefficient, dysfunctional gait," said Jim Bauman, CO, who is in private practice at Edge Homecare Prosthetics and Orthotics in Edison, NJ. "For children with low tone, gait is more staggering and appears clumsy because of what they need to do to get their bodies to move through space."

The role of hypermobility

As noted, hypotonia is often associated with joint hypermobility, which presents its own challenges.

"Hypotonic patients end up with hypermobility, first and foremost in the foot-ankle complex," said Curt Bertram. "These children overpronate right at initial contact, then once they get to midstance, the foot-ankle complex can't stabilize, so their toe lever is shortened and they lose propulsion. With time and repetition in that position, they develop bony modeling that enhances the pronatory deformity."

According to Jason Henry, ligamentous laxity and the associated lack of coordination lead to the kind of balance deficits for which DS patients try to compensate during gait with strategies such as wider stance.

"You see delays in milestones such as cruising," Henry said. (Cruising in this case doesn't refer to muscle cars or girls, but rather the strategy of holding onto furniture or other stabilizing objects while learning to walk.) "You're going to see high guard, a lot of pronation, recurvatum, genu valgum, things like that. It goes all the way up, because hypotonia is global, so you see hyperlordosis, an anterior center of mass, and so forth."

Researchers and clinicians acknowledge that they are somewhat baffled by the connection between hypotonia and hypermobility. Muscles contain motor neurons but ligaments don't—they provide primarily proprioceptive feedback—so the physiological correlation between the conditions isn't clear. Nevertheless, the connection is obvious in the clinic.

"Ligamentous laxity and an associated lack of coordination lead to balance deficits."

"Hypermobile children have greater passive knee ROM but less peak knee flexion than controls."

"Either problem can affect the development of motor skills, but it's even worse if you put them together," said Kathy Martin. "The ligaments are the primary stabilizers of your joints, and if they aren't working well, your muscles are the second line of defense. Kids with low tone don't have either, and I think that's what leads to the delay in the acquisition of motor skills and the gait deviations we see."

A few researchers have investigated hypermobility and gait. In a 2011 study, 29 pediatric patients with hypermobility syndrome had greater passive knee ROM but less peak knee flexion during walking than 37 healthy controls (mean age of participants was 11.5 years). Midstance knee extension during walking was increased in the hypermobile children, but gait speed was not significantly different between groups.[9] Another study found decreased lateral trunk stability during walking in hypermobile children and adults.[10] Research into the hypermobile type of EDS (in this case, in adults), moreover, reported significantly impaired balance and gait; 95% of patients fell during the course of a year.[11] And British researchers have reported that, when joint hypermobility syndrome overlaps with genetic disorders such as EDS and Marfan syndrome, children report relatively high levels of neuromuscular and motor development problems.[12]

Further complicating the picture, however, is that hypermobility can exist by itself, absent the neurological conditions associated with hypotonia. In a study of 8-year-old schoolchildren with generalized joint hypermobility or benign joint hypermobility syndrome, but without other problems, Danish researchers reported that neither condition reduced motor competence or physical activity.[13] A study from the Netherlands reached a similar conclusion.[14]

Even though research into pediatric flexible flatfoot (PFF) is sometimes used as a proxy given the scarcity of data about hypotonia and hypermobility, the clinicians *LER* spoke with for this article generally downplayed the idea of a correlation. Depending on severity and factors such as patient and clinician preferences, most cases of PFF are treated with watchful waiting or simple orthotic strategies. And PFF is not, in any case, a neurological condition.

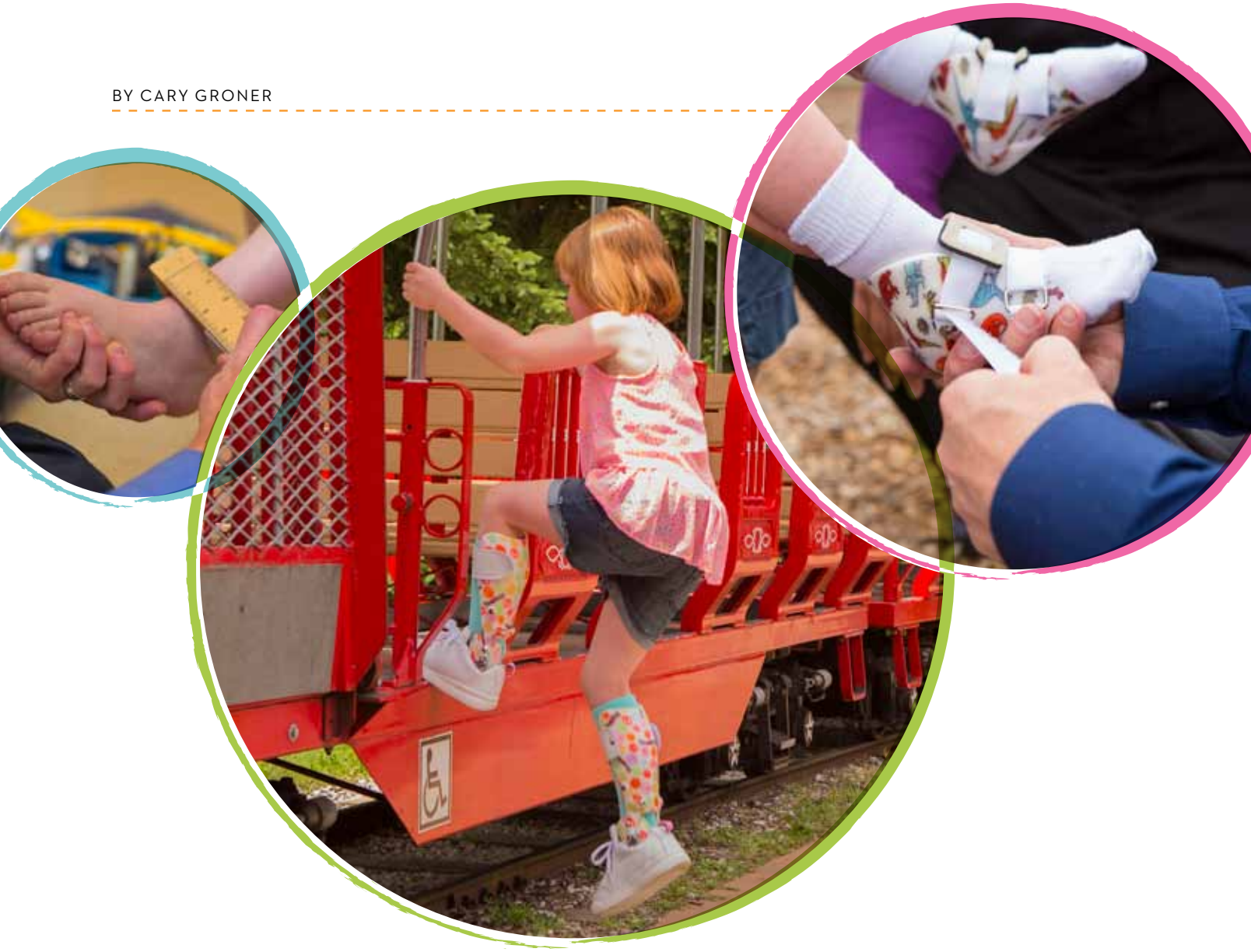
"The problem with Down syndrome children is that they have hypermobility associated with hypotonia, and they end up in a severely overpronated position," explained Curt Bertram. "It's a rotary deformity and very difficult to correct with mild treatment like a foot orthosis. Neither hypotonia nor the associated hypermobility is going to go away in kids with Down syndrome or Prader-Willi syndrome, whereas flexible flatfoot will typically resolve as the child develops and matures."

Clinicians will continue to recognize and treat the manifestations of hypotonia and hypermobility regardless of etiology. That's good news for patients and their parents.

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

References are available at lowerextremityreview.com, or by scanning the QR or tag codes at left.

BY CARY GRONER



Orthotic Solutions for Children with Hypotonia

New research underscores years of positive clinical results

When it comes to orthotic management of pediatric patients with hypotonia, the medical literature is only beginning to document the effectiveness that clinicians have been reporting anecdotally for years.

"The efficacy of treating low tone with orthoses is very poorly studied, but it is the standard of care because I feel intuitively that we can help these kids, and because we've seen good clinical results," said Kathy Martin, PT, DHS, a professor and director of the Doctor of Physical Therapy program at the University of Indianapolis.

Important considerations related to orthotic management of children with hypotonia include which children need what type of device (whether an ankle-foot orthosis, a supramalleolar orthosis [SMO], or an in-shoe foot orthosis) and how early to intervene.

The evidence

There is, at least, some research that examines the case for bracing. Lisa Selby-Silverstein, PT, PhD, coauthored a 2001 paper reporting that foot orthoses affected the gait of children with Down syndrome; effects included reduced heel eversion and transverse plane foot angle during gait, but also a decrease in walking speed.[1] And, in a small (two-subject) 2012 study, researchers found that a flexible SMO improved functional motor performance, whereas a less flexible one impeded it.[2]

Martin published the results of a more robust and compelling study in 2004. Seventeen children with Down syndrome (mean age 5 years, 10 months) were given flexible SMOs and tested three times over 10 weeks on a number of measures that included standing, walking, and running. She found that the devices were associated with significant improve-

ments in postural stability both at the time of fitting (week 3) and after seven weeks of wear (week 10). Martin noted that, the more challenging the task, the more time was needed to see significant improvement, but also that degree of joint laxity did not affect results.[3]

Martin and a couple of colleagues—Julia Looper, PT, PhD, assistant professor of physical therapy at the University of Puget Sound, in Tacoma, WA; and Shelby-Silverstein, who is associate professor of physical therapy at Neumann University in Aston, PA—have put together a proposal for a multisite clinical trial to assess treatment variables, but, even if funding becomes available, results are likely years away.

"Flexible SMOs were associated with improvements in postural stability at the time of fitting and after 7 weeks of wear."

Individual practice

As noted, clinicians have seen results in their own practices.

"When a normal child is learning to walk, their arms come out, their knees are straight, they're moving from side to side, trying to balance," said Curt Bertram, CO, National Orthotic Specialist for Hanger Orthopedic Group who works at Children's Hospital of Wisconsin, in Milwaukee. "That's normal in the prewalking child; but a child with Down syndrome may be in that phase at twenty-eight months, and the condition of their foot isn't going to improve. So in those cases I think it's important to realign the foot in an SMO, to provide a more stable base of support so they can get the proprioceptive feedback they need for balance."

Bertram said that, if the orthosis is flexible enough, it will allow the child to explore movement range but still return them to a more normal biomechanical alignment in the gait cycle.

"I'm in favor of treating as soon as possible because they're already delayed," he continued. "These kids don't have normal biomechanics or muscle tone. We sometimes see older children with Down syndrome who haven't been treated with orthotics, and they typically end up with very deformed, rigid, stiff foot deformities."

The needs of individual patients with hypotonia can be complex and challenging, however.

"We work very closely with the physical therapist in making these decisions," said Jason Henry, MSPT, LO, the practice manager at Hope Orthotics in Spring, TX. "You're looking at age, anatomy, range of motion, strength, coordination, and functional status. I start with an SMO, then work my way up the chain. Are they able to stand independently? If not, they may need a little more support than you get with an SMO. Do they go into a great deal of recurvatum? If so, how are we going to address that?"

Henry prefers flexible SMOs because they allow all the important motions in the sagittal plane—plantar flexion and dorsiflexion at the ankle, forefoot extension, and the like.

"The child can pronate and supinate out of subtalar neutral, which is what you want," he said.

He too believes in bracing earlier rather than later.

"Some people say you shouldn't put braces on kids until they're twenty-four to thirty months old, and I don't agree with that," he said. "They're getting further behind on developmental milestones, and the body is maturing. The limbs are elongating and they're motor learning things in improper alignment, so they'll walk, but what's the quality of the gait?"

PHOTO CONTEST WINNERS

Christian

Meet Christian. Before his SureStep SMOs, this little guy would fall and get discouraged when he tried to walk. Christian now walks around with such pride and confidence that he smiles from ear to ear! He was also the grand prize winner of the SureStep Photo Contest, which put a smile on all our faces.

Kylie

Meet Kylie. Being born on 11/11/11 was only the beginning of this very special little girl's life. At 4 months old she was diagnosed with hydrocephalus, which resulted in a delay in her gross motor skill development. If there's a beat, Kylie is dancing. So, when her physical therapist recommended SureStep SMOs at 17 months, she really stepped up her dance moves. Kylie was the second-place winner in the SureStep Photo Contest.

PHOTO CONTEST WINNERS

Jacob

Meet Jacob. That one extra number 21 chromosome adds a whole lot of unique qualities to this cool kid. Those include hypotonia and pronation. Jacob was shaky and unsure when he first pulled to stand. But with the SureStep SMOs, he gained the stability he needed for freedom on his feet. Jacob has no chance of slowing down any time soon.

Miles

Meet Miles. He is one very motivated young boy. Born with hypotonia, pronation, and PPD-NOS (pervasive developmental disorder not otherwise specified)—one of the three autism spectrum disorders - his mom never thought she'd see the day he would walk. But after wearing his SureStep SMOs, he traded tears for smiles. He is now climbing both steps and ladders with all the confidence a growing young boy should have.

Changes

Faye McNerney, PT, DPT, a pediatric physical therapist in Troy, OH, has seen the field evolve over the course of her 36 years of practice.

"I used to lock up the foot using a rigid SMO or AFO, because I was afraid of what would happen to their foot if they kept rolling inward," she said.

A 2008 study helped quantify the deleterious effects of such approaches, she noted, though it didn't include children with hypotonia. In a British case study of a woman who had her lower leg immobilized following a foot fracture, researchers measured substantial and rapid loss in leg muscle volumes both proximal and distal to the immobilization site. Recovery remained incomplete up to two months after the cast was removed.[4] And, in another study by the same lead author, children immobilized due to hip osteochondritis showed increased ankle stiffness throughout the immobilization period.[5]

McNerney has seen the positive effects of SMOs as her own practice patterns have changed over the years.

"With the flexible braces we immediately see their feet come closer together," she said. "Over time, we don't see the flat-footed gait pattern; a heel-toe pattern develops. In my adult patients with Down syndrome who don't have these flexible SMOs available, I see a much more flat-footed gait pattern."

> "Orthotic interventions have to be carefully selected and matched to the child. Too little is a problem, but so is too much."

When to start

One controversy that has arisen in recent years has to do with the point in the child's development when orthotic intervention is most appropriate. In her studies of Down syndrome patients, for example, Julia Looper has argued that intervening prior to the acquisition of independent walking may interfere with the child's motor-learning skills.[6-8] Kathy Martin acknowledges that this may be the case but points out that it is also important to consider the larger context of cognitive and emotional development.

"We agree that once these kids have learned to walk, orthoses help them," Martin said. "Where we disagree is that Julia's research suggests we should not intervene prior to the acquisition of independent walking. My clinical experience has shown that if you put kids in an orthosis when they are interested in pulling to stand, they start walking sooner. And once they gain independent mobility, their cognitive and social-emotional development explodes. So when I look at a child with Down syndrome who already has cognitive delay, I think the earlier we get them exploring their environment independently, the better their ultimate cognitive function will be."

In her conversation with *LER,* Looper addressed this collegial disagreement by conceding the point.

"She's right," she said of Martin. "My take is that, in kids who are going to be wearing orthoses forever, it doesn't matter that much if they develop control of their ankle. They will be in the orthoses anyway, so exploring their environment and getting the cognitive development is more important."

Looper's position now is that children with milder conditions, who may need to wear orthoses for a shorter period of time, might benefit from watchful waiting.

"I think there is a gradation, and it depends on how much calcaneal eversion we're seeing, and what's going on generally with the child," she said.

Martin emphasized, however, that orthotic interventions have to be carefully selected and matched to the child. Too little is a problem, but so is too much.

"When you and I walk, we pronate and supinate around midline; that's what normal gait is," she said. "Being stuck in pronation throughout the gait cycle isn't normal, but putting a child in a rigid orthosis that holds them in midline constantly isn't normal either. That's where some of the newer SMOs come in; they are more lightweight plastic, thin and flexible and dynamic, so they can bring a child back to midline but not rigidly hold them there. They can pronate and supinate around midline as they walk, and that should be the goal."

Early intervention

If there's a practitioner who has redefined the meaning of early intervention, it's Debbie Strobach, MA, PT, a pediatric physical therapist and splinting specialist at Mercy Children's Hospital in St. Louis, MO.

"We splint children with AFOs before they leave our neonatal intensive care unit," Strobach said.

According to Strobach, preemies born earlier than 30 weeks have a higher risk of hypotonia and muscle imbalance, and may present with significant ankle and hindfoot eversion. This, in turn, causes problems with adjoining muscles.

"We see the fibularis become more powerful than the anterior and posterior tibialis, stretching the medial structures of the foot and putting children at risk for pronation when they're getting ready to stand," she said. "I want them in good alignment so that, as they grow, their tendons and ligaments can support the ankle and foot correctly. That helps the adjoining muscles contract and work as they should, as well."

Strobach and her colleagues use customized solid ankle-foot splints similar to AFOs, in order to counteract such forces. She has been able to assess the effects of her interventions simply by comparing them to children who didn't receive them.

"Kids from other hospitals who weren't splinted in the NICU come to us at nine, twelve, eighteen months of age, and they may even have contractures in the fibularis muscles. We find that the babies can wear splints for maybe a month in the NICU, and then we'll follow them up as outpatients for another month or two. In the NICU, they wear the splints three hours on and three hours off, coordinated with their feeding and handling times. We find that if we splint them early, very few need splints later when they start to stand and walk independently."

Strobach and her colleagues also deal with older children with benign hypotonia, who typically get SMOs or UCBL (University of California Biomechanics Laboratory)-style splints.

"Some of those kids are still not walking by sixteen or eighteen months, so we start them as soon as we get a referral," she said. "Most of them graduate into a plantar orthosis by the time they are four to six years old, and then they're done. The key is having splints that don't inhibit the muscles so you can strengthen them and align the foot correctly throughout the day, for good muscle balance."

Bringing research home

Such approaches, however convincing, remain more a matter of personal clinical experience than evidence-based medicine. Given the paucity of controlled trials, then, clinicians are increasingly documenting their results.

Megan Smith, CO, director of clinical research for SureStep, a maker of flexible SMOs, will present the findings of four recent case studies at the O&P World Congress in Orlando this September.

"The four kids were fifteen or sixteen months old, and presented with diagnoses of developmental delay, benign hypotonia, and significant pronation," she said. "We put them in flexible SMOs and followed them for sixteen weeks, with film every other week."

When the subjects had mastered a skill on the Peabody developmental motor skills scale (e.g., pulling to stand, cruising, taking steps, walking fast), Smith noted the child's age and compared those numbers with the Peabody norms.[9] Although the participants began their evaluations at a baseline of about five months' developmental delay, by the end of the sixteen weeks of study, that disparity had been cut to only a month (see "Orthotic success stories: Four cases in a series," pages 16-23).

"We found that the kids who wore the SMOs had a rate of change 1.8 times greater than normal," she said. "In other words, they were acquiring gross motor skills nearly twice as fast as normal kids and catching up to their peers."

> *"Kids who wore the SMOs had a rate of change 1.8 times greater than normal."*

The future

As clinicians continue to define and refine the effectiveness of their interventions, and if better funding becomes available to conduct randomized trials, ideally the kind of protocols described here will become better documented, adjusted, and, where appropriate, standardized. It will make life easier for practitioners, of course, but the ultimate beneficiaries will be children whose development has been hindered by hypotonia.

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

References are available at lowerextremityreview.com, or by scanning the QR or tag codes at left.

Aniyah

Case Study One

Aniyah was born three months premature, and at the beginning of the study she presented with developmental delay, pronation, and hypotonia. She was prescribed supramalleolar orthoses (SMOs) at 13 months old, at which point she was pulling to stand but not yet taking independent steps. In addition to the SMOs, she received physical therapy once a week for the first eight weeks of the study. Her parents reported that Aniyah wore her SMOs an average of 12 hours a day, six days a week.

On day one of the study, after receiving her SMOs, Aniyah was cruising and taking some steps with hands held, but was very unstable and had a very wide base. Compared to a typical child, her developmental delay was about three months. Four months later, she was very steady while wearing her SMOs (though still unsteady without them), walking with arms down and swinging, and trying to run. She had mastered eight to nine months of gross motor skills in four months, ultimately putting her about one month ahead of her typical peers.

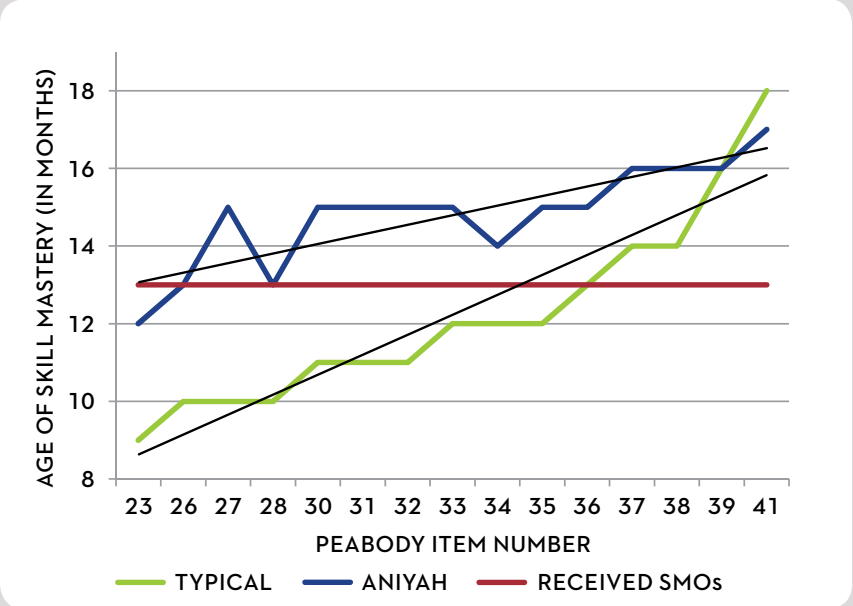
BY MEGAN SMITH, CO

BACKGROUND: Each child in this case series was assessed every other week for 16 weeks to determine mastery of items 23, 26-28, 30-39, and 41 (ranging from "pull to stand" to "walk fast") on the Peabody Developmental Motor Scale. Test instructions were modified as needed for children to understand them. Parents were included in each session and encouraged to play with the child in order to demonstrate the targeted skills. Graphs illustrate age of mastery for each item number for the hypotonic child compared to a "typical" child, with linear trend lines illustrating rate of change, and demonstrate the improved mastery of skills after prescription of supramalleolar orthoses (SMOs). The cases will be presented in September at the O&P World Congress in Orlando, FL.

Day One – Barefoot

Day One – SMOs

Gross Motor Skill Mastery Over Time



PEABODY ITEM DEFINITIONS

23 Pull to stand with support

26 Cruising - 4 steps

27 Lowering to sitting without falling

28 Takes 4 steps with trunk held

30 Standing - child will let go of table and stand for 5 seconds

31 Standing - away from table, child will stand for 3 seconds

32 Stepping - 4 steps with one hand held

33 Standing up from ground without turning more than 20°

34 Walking 8 feet with one hand held

35 Walking 5 steps with no hands held (independent)

36 Standing, picks up toy from ground, stands up and takes 3 steps

37 Creeps up 2 steps

38 Walks 10 feet with narrow base of support, has heel-toe gait for half the distance

39 Creeps down 3 steps backward, without support

41 Walking fast

Source: Folio MK, Fewell R. Peabody developmental motor scales and activity cards. Chicago: Riverside Publishing; 1983.

Colin

Case Study Two

Colin was prescribed supramalleolar orthoses (SMOs) at 16 months old, at which time he was demonstrating pronation, hypotonia, and significant ligamentous laxity. He was pulling to stand but not yet taking independent steps. In addition to the SMOs, he received physical therapy once a week for the duration of the study. His parents reported that Colin wore his SMOs an average of 12 hours a day, seven days a week.

On day one of the study, after receiving his SMOs, Colin was cruising and taking some steps with hands held, but had a very wide base. Compared to a typical child of the same age, his developmental delay was about six months. Four months later, he was walking with a narrow base and low guard, and was stable while walking on grass, uneven surfaces, or transitional surfaces. He had mastered eight to nine months of gross motor skills in four months, ultimately putting him about two months behind his typical peers.

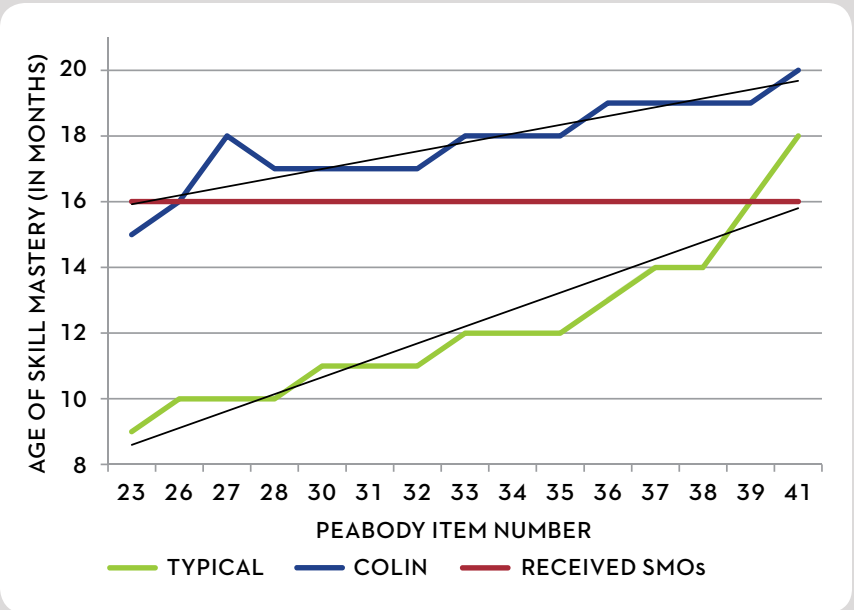
BY MEGAN SMITH, CO

BACKGROUND: Each child in this case series was assessed every other week for 16 weeks to determine mastery of items 23, 26-28, 30-39, and 41 (ranging from "pull to stand" to "walk fast") on the Peabody Developmental Motor Scale. Test instructions were modified as needed for children to understand them. Parents were included in each session and encouraged to play with the child in order to demonstrate the targeted skills. Graphs illustrate age of mastery for each item number for the hypotonic child compared to a "typical" child, with linear trend lines illustrating rate of change, and demonstrate the improved mastery of skills after prescription of supramalleolar orthoses (SMOs). The cases will be presented in September at the O&P World Congress in Orlando, FL.

Day One – Barefoot

Day One – SMOs

Gross Motor Skill Mastery Over Time



PEABODY ITEM DEFINITIONS

23 Pull to stand with support

26 Cruising - 4 steps

27 Lowering to sitting without falling

28 Takes 4 steps with trunk held

30 Standing - child will let go of table and stand for 5 seconds

31 Standing - away from table, child will stand for 3 seconds

32 Stepping - 4 steps with one hand held

33 Standing up from ground without turning more than 20°

34 Walking 8 feet with one hand held

35 Walking 5 steps with no hands held (independent)

36 Standing, picks up toy from ground, stands up and takes 3 steps

37 Creeps up 2 steps

38 Walks 10 feet with narrow base of support, has heel-toe gait for half the distance

39 Creeps down 3 steps backward, without support

41 Walking fast

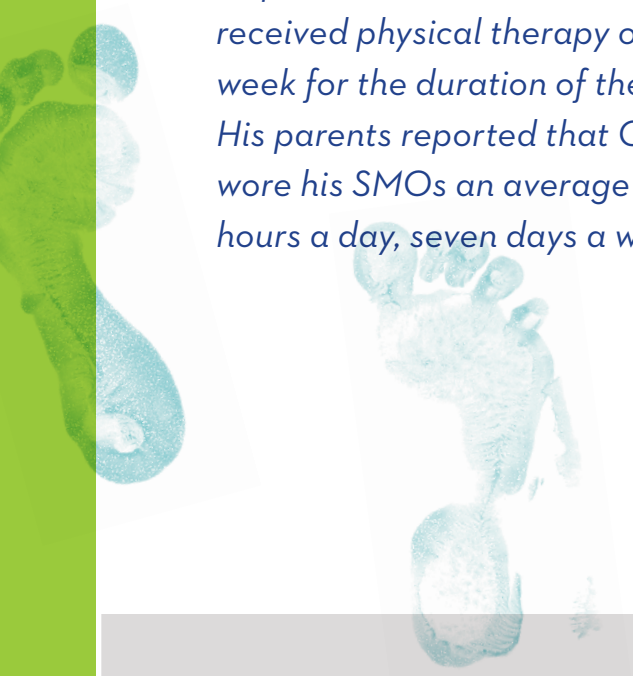
Source: Folio MK, Fewell R. Peabody developmental motor scales and activity cards. Chicago: Riverside Publishing; 1983.

Corwin

Case Study Three

Corwin was prescribed supramalleolar orthoses (SMOs) at 16 months old, at which time he was demonstrating pronation and hypotonia. He was pulling to stand but not yet taking independent steps. In addition to the SMOs, he received physical therapy once a week for the duration of the study. His parents reported that Corwin wore his SMOs an average of 10 hours a day, seven days a week.

On day one of the study, after receiving his SMOs, Corwin was not walking. Compared to a typical child of the same age, his developmental delay was about six months. Four months later, he was walking quickly (almost running) with a narrow base and arms at his sides, and was stable even while carrying objects or while walking on grass or uneven surfaces. He had mastered eight to nine months of gross motor skills in four months, ultimately putting him about two months behind his typical peers.



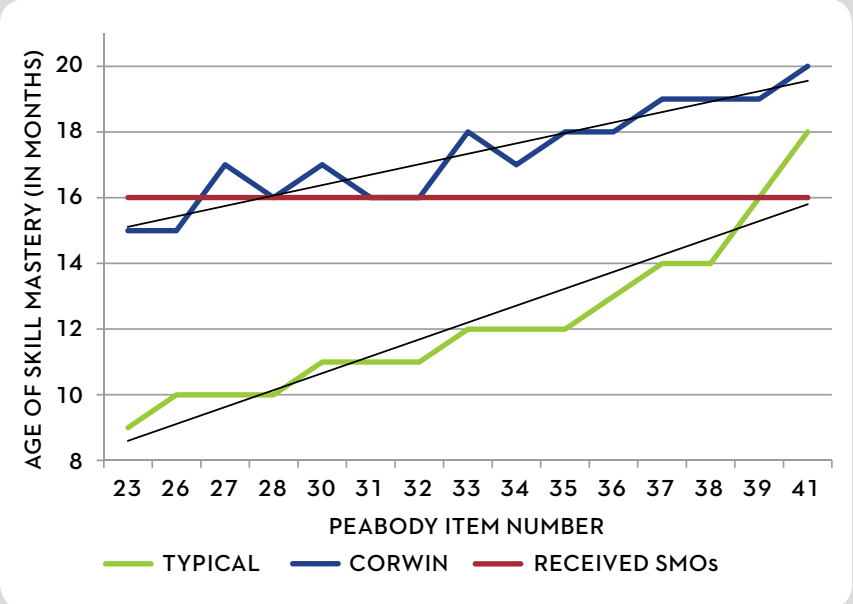
BY MEGAN SMITH, CO

BACKGROUND: Each child in this case series was assessed every other week for 16 weeks to determine mastery of items 23, 26-28, 30-39, and 41 (ranging from "pull to stand" to "walk fast") on the Peabody Developmental Motor Scale. Test instructions were modified as needed for children to understand them. Parents were included in each session and encouraged to play with the child in order to demonstrate the targeted skills. Graphs illustrate age of mastery for each item number for the hypotonic child compared to a "typical" child, with linear trend lines illustrating rate of change, and demonstrate the improved mastery of skills after prescription of supramalleolar orthoses (SMOs). The cases will be presented in September at the O&P World Congress in Orlando, FL.

Day One – Barefoot

Day One – SMOs

Gross Motor Skill Mastery Over Time



PEABODY ITEM DEFINITIONS

23 Pull to stand with support

26 Cruising - 4 steps

27 Lowering to sitting without falling

28 Takes 4 steps with trunk held

30 Standing - child will let go of table and stand for 5 seconds

31 Standing - away from table, child will stand for 3 seconds

32 Stepping - 4 steps with one hand held

33 Standing up from ground without turning more than 20°

34 Walking 8 feet with one hand held

35 Walking 5 steps with no hands held (independent)

36 Standing, picks up toy from ground, stands up and takes 3 steps

37 Creeps up 2 steps

38 Walks 10 feet with narrow base of support, has heel-toe gait for half the distance

39 Creeps down 3 steps backward, without support

41 Walking fast

Source: Folio MK, Fewell R. Peabody developmental motor scales and activity cards. Chicago: Riverside Publishing; 1983.

Hayden

Case Study Four

Hayden was prescribed supramalleolar orthoses (SMOs) at 15 months old, at which time she was demonstrating pronation and hypotonia. She was pulling to stand but not yet taking independent steps, and was very cautious in everything she did. In addition to the SMOs, she received physical therapy once a week for the duration of the study. Her parents reported that Hayden wore her SMOs an average of 12 hours a day, seven days a week.

On day one of the study, after receiving her SMOs, Hayden was not walking. Compared to a typical child of the same age, her developmental delay was about five months. Four months later, she was walking independently and fairly steadily, with a relatively narrow base and low to medium guard. She had mastered eight to nine months of gross motor skills in four months, ultimately putting her about one month behind her typical peers.

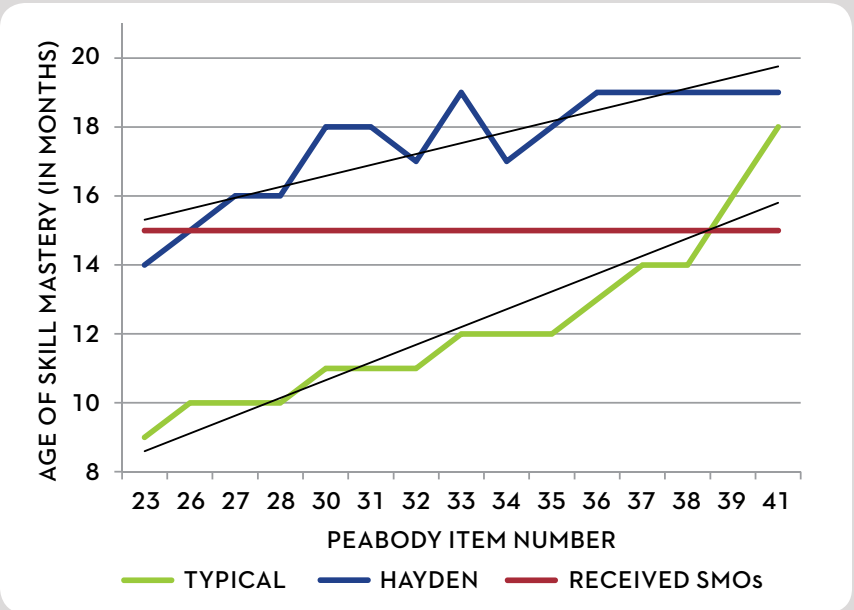
BY MEGAN SMITH, CO

BACKGROUND: Each child in this case series was assessed every other week for 16 weeks to determine mastery of items 23, 26-28, 30-39, and 41 (ranging from "pull to stand" to "walk fast") on the Peabody Developmental Motor Scale. Test instructions were modified as needed for children to understand them. Parents were included in each session and encouraged to play with the child in order to demonstrate the targeted skills. Graphs illustrate age of mastery for each item number for the hypotonic child compared to a "typical" child, with linear trend lines illustrating rate of change, and demonstrate the improved mastery of skills after prescription of supramalleolar orthoses (SMOs). The cases will be presented in September at the O&P World Congress in Orlando, FL.

Day One – Barefoot

Day One – SMOs

Gross Motor Skill Mastery Over Time



PEABODY ITEM DEFINITIONS

23 Pull to stand with support

26 Cruising - 4 steps

27 Lowering to sitting without falling

28 Takes 4 steps with trunk held

30 Standing - child will let go of table and stand for 5 seconds

31 Standing - away from table, child will stand for 3 seconds

32 Stepping - 4 steps with one hand held

33 Standing up from ground without turning more than 20°

34 Walking 8 feet with one hand held

35 Walking 5 steps with no hands held (independent)

36 Standing, picks up toy from ground, stands up and takes 3 steps

37 Creeps up 2 steps

38 Walks 10 feet with narrow base of support, has heel-toe gait for half the distance

39 Creeps down 3 steps backward, without support

41 Walking fast

Source: Folio MK, Fewell R. Peabody developmental motor scales and activity cards. Chicago: Riverside Publishing; 1983.

SureStep℠ C-Fab

C-Fab Service Features:

- Quality fabrication
- Prep option available
- Personalized service
- Quick turn-around time
- Competitive pricing
- Custom to measurements

Ask about our prep option to maximize savings.

Interested in increasing profits?

SureStep is proud to announce the opening of its Central Fabrication division. For more than a decade, SureStep's trained professionals have been providing top-quality components and materials to O&P clinicians across the globe.

By using SureStep C-Fab, not only will practitioners have access to years of knowledge and expertise, but also experience the same quick turn-around times customers are accustomed to with SureStep products. Utilizing SureStep C-Fab will allow O&P professionals to manage profitability, time, and costs easily and effectively.

With a solid reputation for quality and service, SureStep C-Fab is what can help give an O&P practice the edge it needs to stay ahead of the competition.

SureStep℠ academy

online, on demand, in person

1

ONLINE

This course covers the same educational content as the live InServices, but in a webinar format. Designed to last about 3 hours, this course will offer Continuing Education Units (CEUs) for both certified practitioners and physical therapists. CEUs may vary by state.

2

ON DEMAND

Created with convenience in mind, clinicians can take this one-hour course, anytime, anyplace. Specifically-designed for O&P professionals who work with SureStep, this course includes a self-guided presentation including 25 questions that will test users on basic SureStep concepts. Course is approved for 1 CEU.

3

IN PERSON

Certified SureStep presenters travel nationwide teaching about Hypotonia and its effects on gait to physical therapists and O&P professionals. This course is 4.5 hours long, includes dinner and has been approved for 4 CEUs.

SureStep courses are available at no charge.

Check out our website at www.surestepacademy.net for the schedule of courses approved in your state.

You get what you give

When my wife Pam and I started this incredible journey, we did it with passion and faith. SureStep was founded with a passion for improving the lives of children with special needs. And we have always had faith that we will get what we give.

Now, more than a decade later, our core product – the SureStep SMO system - has revolutionized orthotic management for children with hypotonia and has become the mandated method of treatment for this population.

Today, we sell to thousands of practitioners nationwide. Our products are also available in 30 countries worldwide. While we are proud of our achievements, we know that we have much more work to do.

We continue to invent and develop products and offer services that keep us focused on our core mission. You'll see in this catalog that we are introducing three new products: the DA Hinge, HEKO PreFab, and our Criss-Crossers. We have recently become distributors for the Allard KiddieGAIT system and are launching two new divisions of our organization: SureStep Academy, our educational division, and SureStep C-Fab, our central fabrication division.

It is my hope that by satisfying our customers with these types of innovative technology, superior quality, value, and service, we will solidify our reputation and commitment to this field. When these attributes are combined with the performance of our products, I believe SureStep can become an incredibly powerful force to make a difference in this world.

We share that same commitment to our employees. We continue to contribute to building and sustaining a safe, inclusive, engaged, and socially responsible workplace focused on delivering life-changing results. For me, the success of our work has evolved seamlessly. It has reinforced that as the CEO of SureStep, leadership is a form of service. It also reinforces the true power of giving back.

I hope that your belief in the effectiveness of our products evolves into a mission to change the lives of children along with me. We had faith in our company's success, and we continue to believe that if we act with uncompromising honesty and integrity in everything we do, we will get what we give.

SINCERELY,
BERNIE VELDMAN, CO
CEO, SURESTEP

SureStep℠

SureStep Products

SureStep SMO

INDICATIONS

- Pronation
- Low muscle tone (hypotonia)
- Developmental delay
- Delay in acquiring gross motor skills
- Poor coordination or balance

CONTRAINDICATIONS

- High muscle tone
- Spasticity
- Tight heel cords
- Tight peroneals

SUGGESTED CODES

- L1907 x1
- L2275 x2

CUSTOM FABRICATED TO MEASUREMENTS

Casts and measurements required when #5 measurement exceeds 2 3/4"

SureStep SMO

The SureStep SMO remains the most advanced method of controlling excessive pronation and providing stability to the hypotonic population. Suitable for patients up to 80 lbs.

Through the use of extremely thin, flexible thermoplastic, the SureStep SMO compresses the soft tissue of the foot. This compressive force stabilizes the foot and ankle complex while still allowing for the development of the intrinsic muscles, muscle strategies and movement patterns necessary to develop a normal, natural gait pattern.

The SureStep SMO has revolutionized orthotic management for children with hypotonia and has become the mandated method of treating this population in many areas of the world.

No Casting Required

Many custom orthoses require casting which is a time-consuming, messy and often a traumatic process for smaller children. SureStep SMOs require only nine easy measurements. It's simple, it's quick, and it's effective.

No Adjustments, No Waiting

SureStep products are available to orthotic and prosthetic facilities nationwide and in 30 countries around the world. Once we receive an SMO order from a certified O&P clinician, the product will be fabricated and shipped within 2 business days. No returning for adjustments, no more waiting for long fabrication times. That means a child can be wearing their SureStep SMOs within a week.

Want to find a clinician in your area? Email or call us with your zip code and we'll give you a list of facilities nearest you. You can reach us at:

info@surestep.net or 877-462-0711

Benefits of SureStep SMOs

- Improved efficiency
- Increased stability
- Enhanced alignment

Visit our site for more information and videos | **SureStep.net**

SURESTEP PATTERNS

You know you are getting SureStep brand and quality when you see our exclusive patterns. Visit our website to choose your favorite!

LIVE OUTSIDE THE U.S.?

SureStep products are available in many areas of the world. Our list is growing daily. To learn more about how you can obtain our products internationally, please email us your request at info@surestep.net.

FOLLOW US

About Our Trimlines

The SureStep SMOs are carefully marked for "right" and "left" and although they may look different than other orthoses, our unique patented trimlines are an integral part of the system that enhances a child's ability to run, jump and play naturally.

The "lateral", or outside trimline of the SureStep SMO extends further than the "medial", or inside trimline. This is in sharp contrast to most traditional orthoses.

Enhanced Alignment

Excessive pronation puts undue stress on the knees and hips by changing the alignment of the lower extremities, taxing the muscles of the legs greatly.

This often results in complaints of fatigue or "tired legs". SureStep helps to stabilize the foot and ankle relieving stress and allowing the muscles to work more efficiently.

BigShot/BigShot Lite

INDICATIONS

- Pronation
- Hypotonia
- Triplanar instability in weight bearing
- Inability to stand independently
- Mild toe-walking

CONTRAINDICATIONS

- High muscle tone
- Spasticity
- Severe toe-walking

SUGGESTED CODING

- L1907
- L2280
- L2275x2

CUSTOM FABRICATED TO MEASUREMENTS

Casts and measurements required when #5 measurement exceeds 2 3/4"

SureStep BigShot

A growing child means eventually outgrowing the original SureStep SMO. The BigShot and BigShot Lite are the perfect solutions for older children who still need the stability of SureStep.

The SureStep BigShot incorporates all of the same features of the SureStep SMO with the addition of a soft silicone-like inner boot to ensure comfort. The flexible, plastic outer frame serves to provide the compression and stability that children exceeding 80 lbs. need, but allow all of the normal, necessary flexibility for a smooth, natural gait pattern.

SureStep BigShot Lite

The BigShot also comes in a "lite" version for children between 50 lbs. and 90 lbs. The BigShot Lite offers a thinner inner boot, as well as a thinner, more flexible plastic than its stronger counterpart, the BigShot.

Want to find a clinician in your area? Email or call us with your zip code and we'll give you a list of facilities nearest you. You can reach us at:

info@surestep.net or 877-462-0711

Indy 2 Stage

Visit our site for more information and videos | **SureStep.net**

SureStep Indy 2 Stage

A uniquely designed orthosis developed to help children reach their potential. This exceptional "orthosis within an orthosis" allows for the SureStep SMO to be utilized independent of the AFO. Children can work through a variety of transitional skills without impeding normal muscle function.

When used together, the SureStep SMO locks securely into the AFO transforming this truly dynamic SMO into an AFO that provides triplanar stability without restricting normal usage of intrinsic musculature of the foot. By doing this, children can continue to develop those intrinsic muscles, muscle strategies and movement patterns necessary for an improved gait pattern.

Product Benefits

Therapists can use the SureStep Indy 2 Stage as a tool for children as they transition through their upright gross motor skill development. The Indy 2 Stage is ideal for children who require the proximal support of an AFO to achieve independent stance, but still benefit from an SMO for proper crawling, pull-to-stand and cruising activities.

It is also a great solution for children as they progress to taking independent steps with the AFO. It allows for them to continue to work on improving motor plans with the internal SureStep SMO, thus providing a smooth transition to less bracing.

Battling Fatigue

Children with disorders that cause increased fatigue throughout the day can begin their day in only the SureStep SMO. This will serve to facilitate increased effort to maintain their functional muscle groups. As they fatigue throughout the day, the external AFO can be added to maintain a stable and functional gait pattern.

This product is suitable for patients up to 100 lbs.

INDICATIONS

- Low muscle tone (hypotonia)
- High muscle tone (hypertonia)
- Flexible pronation or supination
- Poor proprioceptive awareness
- Difficulty with transitional skills
- Sagittal and/or frontal plane weakness or instability

CONTRAINDICATIONS

- Significant ankle contractures
- Severe spastic involvement

SUGGESTED CODING

- L1960 (Solid)
- L1970 (Hinged)
- L2200 (Hinged)
- L2275x2
- L2280

VARIATIONS

- Articulated (comes standard with Dual Adjustable Joint)
- Solid ankle
- Posterior leaf spring

CUSTOM FABRICATED TO MEASUREMENTS

Casts and measurements required when #5 measurement exceeds 2 1/2"

Pullover & Advanced

PULLOVER

INDICATIONS
- Pronation
- Low muscle tone (hypotonia)
- Mild-hemiplegia
- Drop foot
- Weakened dorsiflexors
- Sagittal plane instability

CONTRAINDICATIONS
- Significant spasticity
- Tight heel cords
- Fixed deformities of the foot
- Tight peroneals

SUGGESTED CODING
- L1970
- L2275
- L2280
- L2210x2

CUSTOM FABRICATED TO MEASUREMENTS
Casts and measurements required when #5 measurement exceeds 2 1/2"

ADVANCED

INDICATIONS
- Pronation
- Low muscle tone (hypotonia)
- Joint hypermobility
- Delayed standing
- Inability to stand for prolonged times

CONTRAINDICATIONS
- Strong spasticity
- Significant contractures
- Significant foot deformities

SUGGESTED CODING
- L1960
- L2275x2

VARIATIONS
- Full foot plate
- SureStep trimlines

CUSTOM FABRICATED TO MEASUREMENTS
Casts and measurements required when #5 measurement exceeds 2 1/2"

SureStep Pullover

The SureStep PullOver is an amazing tool that not only facilitates improved stability of the foot and ankle complex using a SureStep SMO, but it is also the only SMO that incorporates a true dorsiflexion assist through the use of a removable proximal strut. This allows for the use of the device as a SureStep SMO, whenever proximal support or dorsiflexion assist is not necessary. Suitable for patients up to 80 lbs.

When appropriate, the proximal strut can be attached easily and the PullOver becomes a free-motion, dorsiflexion assist AFO. The PullOver is a perfect solution for many children with mild hemiplegia, or for children with hypotonia that need just a little bit of additional help into dorsiflexion.

SureStep Advanced

The SureStep Advanced AFO is the ideal device for pre-walkers. The SureStep Advanced incorporates the same concept of circumferential compression as the SureStep SMO. Plus, it extends to full AFO height, integrating increased sagittal plane stability to help children find their ideal standing position. Suitable for patients up to 80 lbs.

While the SureStep Advanced AFO prevents plantarflexion, simple changes to the proximal strapping configuration allows for varying degrees of dorsiflexion.

The SureStep Advanced AFO is very adaptive. When children are ready for ambulation, it can be converted easily to a SureStep SMO. This unique device can be fit with either a full footplate or a modified SureStep footplate.

HEKO & HEKO PreFab

Visit our site for more information and videos | **SureStep.net**

SureStep HEKO Custom

SureStep HEKO is the first and only pediatric hyper extension knee orthosis to incorporate a 4-axis knee hinge, for smooth, anatomically correct flexion and extension. This exceptional device provides localized control of the knee, preventing hyperextension, valgus and varus, while allowing full flexion and extension. With adjustable extension stops, the HEKO offers up to 30 degrees of adjustability. Each SureStep HEKO is custom fabricated to measurements using SureStep's CAD/CAM system, but can be fabricated from a cast impression or scan. The HEKO comes complete with 2 anti-migration/suspension sleeves.

With two hinge sizes, the SureStep HEKO is ideal for patients up to 100 lbs.

SureStep HEKO PreFab

The HEKO PreFab incorporates all of the features of the original HEKO into a prefabricated version.

Integrating the same unique 4-axis knee hinge, this more flexible HEKO PreFab allows even more adjustability.

HEKO PreFab provides the physical therapist or O&P professional a prefabricated option that can create much greater knee stability and improve function dramatically.

Available in 4 prefabricated sizes, in either soft purple or black, HEKO PreFab is idea for patients up to 50 lbs.

INDICATIONS

- Knee instability
- Genu-recurvatum
- Flexible Genu-valgum or Genu-varum

CONTRAINDICATIONS

- Fluctuating edema

SUGGESTED CODING

- L1846 (Custom)
- L1845 (PreFab)
- L2820
- L2830
- L2750x2
- L2397 (suspension sleeve)

CUSTOM HEKO FROM MEASUREMENTS

PREFAB SIZES
S, M, L, XL

HINGES CAN BE SOLD SEPARATELY

Criss Crossers & De-Rotation Straps

SureStep Criss Crossers

The first and only device developed to discourage w-sitting, Criss Crossers use a unique audio cue to remind children to change their position.

Available in 5 standard sizes, this innovative design can be worn under most regular clothing and will fit most infants, and small children. The small sealed battery (similar to a watch battery) and tone generator easily unplug to allow normal cleaning and care.

Criss Crossers are a simple and effective method of discouraging w-sitting without putting any other positional limitations on children.

DEROTATION STRAPS

INDICATIONS

- Flexible internal or external femoral or tibial rotation

CONTRAINDICATIONS

- Fixed rotational deformities

SUGGESTED CODING

- L2999

PEDIATRIC & ADULT SIZES AVAILABLE

FOLLOW US

SureStep De-Rotation Straps

The SureStep De-Rotation Straps offer dynamic control for mild femoral or tibial rotation. The latex-free elastic hook-and-loop strap system easily attaches to shoelaces or to lower extremity orthoses. And its low-profile design easily hides under clothing.

Available in both beige and white, the universal "pediatric" size will fit patients up to 5 ft. tall. Also available in a universal "adult" size, these devices can be custom-fitted to the patient simply and quickly.

This product gives O&P professionals and physical therapists an easy-to-use solution for flexible internal or external femoral or tibial rotation.

TLSO

Visit our site for more information and videos | **SureStep.net**

SureStep TLSO

The SureStep TLSO has redefined spinal management. The soft, flexible plastic serves well to create improved upright positioning, while still allowing for slight movement in all planes.

By more evenly distributing pressure circumferentially, this unique device creates stability without rigidity, permitting the core to continue to strengthen. Custom fabricated from measurements, cast impressions or scans, the SureStep TLSO comes complete with a soft interface which is available in multiple thicknesses for improved comfort and compliance.

The SureStep TLSO is available with either an anterior or posterior opening and can be modified to accommodate G-tubes, baclophen pumps, etc. The SureStep TLSO is the ideal solution for many wheelchair bound children who have difficulty in maintaining an upright sitting position.

Breathing Modifications

Anterior Window: This simple, yet effective modification facilitates anterior expansion of the ribs and abdominal areas. The anterior window requires a posterior opening and is a perfect solution for children who tend to breathe lower.

Gill Modification: While this modification may look small, its impact will be great. The gill modification simply allows the chest to expand laterally and enables improved breathing within the TLSO. This modification can be added to either an anterior or posterior opening TLSO, and can be added at the time of fabrication, or anytime thereafter.

Anterior Window

Gill Modification

INDICATIONS

- Inability to control position in wheelchair
- Postural instabilities of the trunk
- Flexible kyphosis or lordosis

CONTRAINDICATIONS

- Fixed scoliotic curvatures
- Adolescent idiopathic scoliosis
- Severe respiratory limitations

SUGGESTED CODING

- L0482x1

VARIATIONS

- Anterior opening
- Posterior opening
- Early Intervention (.75mm)
- Standard (1.25mm)
- Adult (2mm)

CUSTOM FABRICATED TO MEASUREMENTS, CASTS OR SCANS

SURESTEP PATTERNS

You know you are getting SureStep brand and quality when you see our exclusive patterns. Visit our website to choose your favorite!

www.surestep.net

Shoes/Sandals

SureStep Children's Footwear

SureStep shoes have been custom designed specifically for children who wear orthoses. Their wider, deeper heel, toe box and instep allow for adequate room and a comfortable fit. The unique tread promotes intrinsic movement and flexibility while a special "cut-line" allows for easy shoe modifications.

SHOES

- Decreased number of eyelets for easier donning and doffing
- High quality upper and solid sole with pink or blue trims
- Sole designed for custom modification
- Two removable insoles for perfect fit
- Low profile and highly functional
- **Sizes: Toddler 3 to Youth 12**

SANDALS

- Sandals are designed with more height and longer straps to accommodate various types of orthoses
- Same flexible sole to allow intrinsic movement
- Highly functional, durable material in pink or black
- Sole designed for custom modification
- **Sizes: Toddler 3 to Youth 12**

Components

Visit our site for more information and videos | **SureStep.net**

SureStep HEKO Hinge

Originally designed and developed for use in the SureStep HEKO, this unique hinge is the only 4-axis pediatric hinge available. Perfect for use in KAFOs, knee orthoses and elbow orthoses; it is lightweight and low profile. The hinge has adjustable extension stops that allow specific range of motion limitations and adjustability. Available in two sizes, the HEKO hinge will work well for patients up to 100 lbs.

SureStep DA Hinge

Available in both toddler and youth sizes, the SureStep DA Hinge does what other hinges cannot. The tall proximal upright extends well into the AFO strut, facilitating corrugation of the plastic and strengthening the strut. This allows AFOs to be fabricated with much thinner, lighter weight plastics. The inverted "Y" distal stirrup connection allows for a solid, stable attachment to a molded footplate, further strengthening the AFO. Adjustments for both dorsiflexion and plantarflexion stops allow the O&P clinician to adjust for as much, or as little ROM as necessary. Dorsiflexion assist bands can be easily added at the time of fabrication or anytime thereafter. At less than 3/8″ thick, the DA Hinge is lower profile than most poly hinges.

SureStop

The SureStop is a removable, adjustable plantarflexion stop that not only reduces the noise normally associated with plastic plantarflexion stops, but also allows for adjustment to varying degrees of plantarflexion. The SureStop eliminates the need for gluing and dealing with lost stops. It locks securely in place with small "teeth" that grip the plastic, and allows the SureStop to click right in.

SureStep Free Motion Hinge

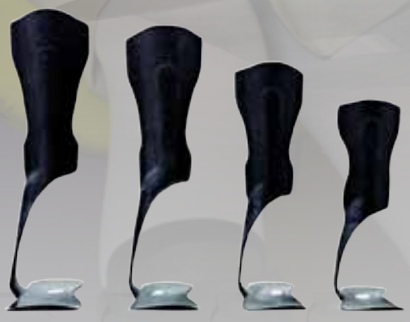
The SureStep Free Motion Hinge is a simple yet elegant hinge that is quickly becoming the first choice by O&P clinicians across the country. Boasting the lowest profile of any hinge in its class, the SureStep Free Motion Hinge is quick and easy to use in fabrication. With no alignment rods or jigs necessary, these hinges can be visually aligned, formed and pulled within a few short minutes, saving valuable fabrication time. Fabrication technicians love this hinge for its ease of use. Practitioners love it for its low profile and cost savings.

SureStep Dorsiflexion Assist Band

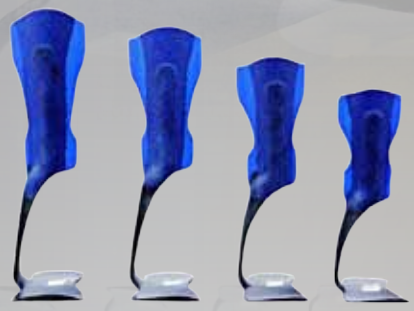
Simplicity strikes again. This small band can be added to most new or existing AFOs to add dorsiflexion or plantarflexion assist. The smooth even pull that it generates is perfect for both small and large AFOs. Tension can be adjusted by simply changing the attachment point, and at less than 1/8" thick, bulk will never be an issue.

SureStep℠

Official Distributor of
KiddieGAIT™ & KiddieROCKER™



KiddieGAIT™
Sizes available to fit infants to approximately 10 years.



KiddieROCKER™
Sizes available to fit approximately 5 to 10 years.

SureStep is a proud distributor of the Allard KiddieGAIT™ and KiddieRocker™. The SureStep SMO and Allard Kiddie product line have always made a great orthotic combination, so it only makes sense to offer them together. Because of our partnership, SureStep customers can easily order both product lines at one convenient time. Give us a call to discuss our competitive pricing.

Whether you order one or a dozen, we know you'll be happy with the results.

SureStep℠
www.surestep.net

allard USA
www.allardusa.com