Clinical Considerations in Cerebral Palsy and Spasticity

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ABSTRACT

The ultimate goal for management of patients with cerebral palsy is to help them grow up to become as independent as possible, learn to make their own choices in life, and pursue their own dreams. Optimal mobility is crucial to achieving independence and is also necessary for better health and quality of life in these patients. This article discusses the treatment of spasticity in cerebral palsy, addresses tone management issues in relationship to mobility and physical fitness, and introduces the reader to a comprehensive approach to the management of patients with cerebral palsy. (*J Child Neurol* 2001;16:10–15).

This article is not intended for the practitioner who is in search of a simple algorithm for managing spasticity in the child with cerebral palsy. Neither is it addressed to those physicians who believe that the management of the child with cerebral palsy begins and ends with the treatment of spasticity. The goals of this article are (1) to bring a new perspective to "spasticity" and its effects on mobility in patients with cerebral palsy and (2) to guide the practitioner toward a comprehensive, goal-directed approach to the patient with cerebral palsy that optimizes each patient's chances for mobility and independence.

Cerebral palsy is typically classified into spastic, dyskinetic, ataxic, and mixed varieties.¹ Although the majority of patients with cerebral palsy (>85%) are classified as having the "spastic" variety, there are very few patients with "pure" spasticity.² Moreover, the defining feature of cerebral palsy is not spasticity but rather the patient's inability to control movements and posture. Reducing tone does not correct the underlying disorder of motor control. Additionally, children with cerebral palsy are at high risk for many other associated impairments, including visual disturbances, hearing loss, speech and language difficulties, epilepsy, nutritional disturbances, learning difficulties, cognitive impairments, depression, and behavioral problems. These associated disorders deserve equal if not greater attention than management of the child's spasticity if we are to help each patient with cerebral palsy succeed in life.

At our Cerebral Palsy Center, we offer a unique, proactive approach to our patients (child, adolescent, and adult) that is optimistic, goal directed, and comprehensive. We target four main areas of need: (1) communication and education, (2) mobility, (3) physical fitness, and (4) independence. Maximal independence in life is the desired outcome for every child. It is noteworthy that "spasticity" is not listed as one of our main focus areas. That is because we consider spasticity and tone management in the context of several other important factors that affect a patient's mobility (Figure 1). Additionally, although this article will focus primarily on spasticity and mobility in cerebral palsy, management of every child with cerebral palsy should begin with an assessment of their communication and educational needs.

MOBILITY

Mobility is crucial to the long-term health and overall wellbeing of patients with cerebral palsy. Without mobility, these patients face severe and substantial risks of chronic pain³ and poor health due to sedentary lifestyle, including diseases such as osteopenia/osteoporosis,⁴ scoliosis, hip fracture, heart disease, and obesity. Immobility also leads to dependence and a loss of freedom. Dependent patients are at high risk of depression due to feelings of hopelessness,

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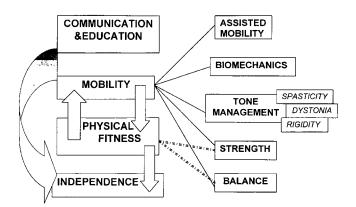


Figure 1. Spasticity, mobility, and the management of cerebral palsy. Spasticity limits mobility. Tone management is one aspect of a comprehensive approach to the management of patients with cerebral palsy. This schematic depicts the relationship between spasticity and other factors that affect mobility and independence in patients with cerebral palsy.

helplessness, and their perceived lack of control over their own life choices. To optimize each patient's mobility, the physician must address several factors, including biomechanics, tone, strength, balance, and physical fitness.

BIOMECHANICS

Optimal biomechanics serve to (1) promote normal spine, bone, and joint development; (2) optimize range of motion and muscle/tendon length as the child grows; and (3) prevent deformities of the limbs and spine that can further compromise a patient's functional ability and health. Several different approaches are available for optimizing biomechanics (Table 1).

Stretching helps maintain and improve muscle-tendon length and joint range of motion. Standing and weight-bearing exercise promote normal development of the hip, spine, and bones.⁵ Orthotics and bracing can be used to improve gait and prevent muscle contractures and progressive limb deformity, particularly during phases of rapid growth.^{6,7} Serial casting and botulinum toxin injections are both effective in improving range of motion at contracted joints.⁸⁻¹⁰ Finally, despite the increasing use of nonsurgical interventions to promote optimal biomechanics in cerebral palsy, the judicious use of orthopedic surgery remains an important treatment modality in these patients.¹¹ In any given patient with cerebral palsy, it is important to carefully weigh the risks versus benefits of each modality and choose the best strat-

Table 1. Strategies for Optimizing Biomechanics

Stretching Weight bearing (standing) Orthotics and bracing Serial casting Botulinum toxin Orthopedic surgery egy. For example, consider the following different paradigms for managing contractures of the heel cords or hamstrings (Tables 2 and 3).

Toe Walkers

Both of the children with heel cord contractures (see Table 2) need orthotics to improve their gait and prevent progressive deformities of the foot. However, patient A may respond to ankle-foot orthoses alone, whereas patient B is likely to require additional interventions. In patient A, a good first strategy would be to try a dynamic or hinged ankle-foot orthosis that blocks plantar flexion but permits her to continue to dorsiflex her foot. The same dynamic ankle-foot orthosis can be worn at night as a stretching splint. This approach will allow her to continue to strengthen her tibialis anterior during gait while stretching her gastrocnemius and heel cord. She may eventually be able to wean to less bracing. A less suitable strategy for patient A would be to use rigid ankle-foot orthoses. Although they will effectively prevent progression of her heel cord contractures, the rigid ankle-foot orthoses will also weaken her ability to dorsiflex her foot and may result in undesired alternative gait strategies.¹²

In contrast, patient B will most likely fail a trial of orthotics unless something else is done first to increase the range at her heel cords. In her current state, she will not be able to get her heel down in an appropriate orthotic, and forcing her to wear an ankle-foot orthosis may destroy the orthotic and/or result in skin breakdown and foot pain. Stretching alone will also likely prove to be a frustrating experience for both the child and the parent and yield little benefit. She could be managed with serial casting of both lower legs for 4 to 6 weeks prior to placement in ankle-foot orthoses. Another, equally viable option is to use botulinum toxin to weaken her gastrocnemius muscles, followed by a briefer round of serial casting (2-4 weeks), before placing her in dynamic ankle-foot orthoses. The advantage of using botulinum toxin in patient B is that it can reduce the length of time required for casting, and the combination of botulinum toxin and casting can produce a more rapid effect than serial casting alone.

Scissoring and Crouch Gait

Both patients described in Table 3 have appropriate orthotics for the lower leg and only mild limitation of range at the adductors. The scissoring of the legs observed in patient C is due to shortening of the medial hamstrings. Patient C is a small child. Therefore, it is reasonable to treat him with botulinum toxin injections to the medial hamstrings, followed by serial casting (long-leg casts) for 3 to 4 weeks. Repeat rounds of botulinum toxin and serial casting may be needed. Surgical release of the hamstrings should be considered if other treatments fail or if there is evidence of progressive lateral migration of the femoral head. However, hamstring contractures can recur, and the patient may have to undergo a repeat lengthening procedure by the time he finishes growing.

	Patient A	Patient B
Age	5 yr	5 yr
Diagnosis	Spastic diplegia	Spastic diplegia
Assistive device	None; patient walks unassisted	Patient holds onto the wall or parents for support
Gait	Toe walker, no heel strike	Toe walker, no heel strike
Range of motion* (ankle)	(+5° bilaterally); despite stiffness, patient's ankle can be passively brought past neutral	(-10° bilaterally); moderate heel cord contractures, cannot bring ankle to neutral
Active foot dorsiflexion	Can bring heels to floor during stance Mild dorsiflexion of toes and forefoot on command	No voluntary dorsiflexion Paradoxical plantar flexion of the foot occurs when patient tries to pull up her toes

* Passive range of motion at the ankle is measured with the patient supine and knees fully extended. The numeric values listed refer to the extent of dorsiflexion relative to neutral. (Neutral = 0°; neutral is achieved when the ankle is brought to 90°.)

Patient D is a large adolescent and probably will not tolerate lengthy periods of bilateral long-leg casting. Serial casting will severely limit his ability to move and will make personal hygiene difficult. Moreover, serial casting will weaken his quadriceps muscles, which will further worsen his crouch gait. In our experience, botulinum toxin is relatively ineffective for treating hamstring contractures in larger children and adolescents. Due to the size of the muscles involved and dose limitations of the drug, it is extremely difficult to inject enough botulinum toxin to sufficiently weaken the hamstrings in these patients. On the other hand, tendon releases are an excellent option in teenagers with moderate to severe contractures of the hamstrings and crouch gait, and it is likely that these patients will not require a repeat lengthening procedure since they are essentially finished growing. Care must be taken to minimize the period of postoperative immobility and to encourage the patient to resume weight bearing as soon as it is safe to do so.

A good rule to remember before recommending serial casting or any brace/orthosis is that the longer and harder you immobilize a joint, the weaker the muscles surrounding that joint will become. For example, hip-knee-foot orthoses can be used to prevent a patient from crouching at the knees and ankles. However, bracing the entire leg results in progressive atrophy and weakness of the quadriceps and lower leg muscles. This approach ultimately worsens the crouch gait, leaves the child in pain or dependent on the heavy

braces indefinitely, and can greatly limit his mobility. Even transient immobilization from serial casting or following surgery will weaken leg muscles. Thus, it is important to use only that bracing that is necessary to prevent or correct deformity, to continually re-evaluate a patient's need for orthotics, and to use adjunctive strengthening and strengthening exercises to maintain and improve function.

SPASTICITY

The goals for treating spasticity in patients with cerebral palsy are threefold: (1) to improve range of motion, (2) to ease the patient's ability to move, and (3) to make the patient more comfortable.

Before we can effectively manage spasticity, it is important to clearly define what we mean by this term. In the strictest sense, spasticity is defined as an increased resistance to passive stretch that is velocity dependent. However, the term "spasticity" is used inconsistently and means different things to different people. Spasticity is frequently used to denote any stiffness or increased tone in patients who have other evidence of corticospinal tract involvement (eg, hyperreflexia, extensor plantar responses). As a result, describing a patient with cerebral palsy as "spastic" tells us that the patient is stiff but gives us very little information as to the types of stiffness or the potential mechanisms underlying that stiffness. Although parceling out the stiffness

Table 3.	Comparing Two Patients With Hamstring Contractures
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	Patient C	Patient D
Age	4 yr	17 yr
Diagnosis	Spastic diplegia	Spastic diplegia
Assistive device	Reverse walker	Forearm crutches
Gait	Scissoring steps	Severe crouch gait
Range of motion* (knee) Active knee extension	(120°) moderately severe contractures bilaterally Able to achieve full knee extension during stance	(90–100°) severe contractures bilaterally Crouched in stance, cannot fully extend knee

*Passive range of motion at the knee is measured at the popliteal angle with the patient supine with one leg raised (hip at 90°).

into different subtypes in these patients is difficult, it is important to do if we are to arrive at effective treatments for tone management.

As one who has a unique perspective on this subject, I offer the following description of what it feels like to live inside a "spastic" body, with the added caveat that other patients may provide different descriptions of their stiffness:

In the conscious, nonanesthetized, nonmedicated state, I always feels stiff, regardless of my position (supine, sitting, standing, walking). This stiffness is not constant in intensity; it fluctuates widely from day to day and even from moment to moment; it can be exacerbated by environmental and emotional factors such as cold temperatures and sleep deprivation. This stiffness can increase suddenly and without warning or increase more gradually as a progressive tightening of the muscles from my feet to my legs to my trunk, arms, and neck. It feels like my muscles are in a 'tug of war,' and it is exceedingly difficult or even impossible to 'will' myself to relax. In fact, the effort of trying to relax often makes things worse.

It is my opinion that a better term for the stiffness in patients with cerebral palsy is "hypertonicity." This term encompasses the broad range of tone abnormalities experienced by these patients, including spasticity, dystonia, and rigidity. Dystonia manifesting as progressive flexion deformities of the wrist and elbow is common in patients with spastic quadriplegia and hemiplegia. Rigidity also occurs in patients with cerebral palsy when they are excited or stressed, after which their limbs become unbendable for a time. The brain injury in patients with spastic cerebral palsy is likely to involve more than one motor system including pyramidal (corticospinal tract injury in periventricular leukomalacia) and nonpyramidal (eg, developing basal ganglia) pathways.^{2,13}

Management of hypertonicity in the patient with cerebral palsy involves multiple approaches, including environmental manipulations such as reducing sleep deprivation, alleviating pain, therapeutic use of heat and/or deep tissue massage, and reducing prolonged periods of immobility.14 Multiple oral medications are used to treat "spasticity" but have limited efficacy in most patients due to unacceptable side effects.¹⁵ Oral medications to treat dystonia, such as levodopa, can improve motor function in patients with spastic quadriplegia¹³ or mixed athetoid/spastic forms of cerebral palsy.¹⁶ Neurosurgical interventions include the baclofen pump and selective dorsal rhizotomy. Placement of a pump to allow the delivery of baclofen directly to the spinal cord is more effective at reducing spasticity and dystonia without the cognitive side effects that are frequently seen with oral administration of the drug.¹⁷ Selective dorsal rhizotomy involves the selective ablation of dorsal nerve rootlets and results in reduced tone in the lower extremities.^{18,19} These surgical interventions offer dramatic tone reduction for the patient with cerebral palsy, but it is important to be sure that the patient and family understand the potential risks and side effects of these procedures.

Management of "spasticity" is difficult. Patients and their families often find themselves choosing between living with the spasticity or dealing with the unacceptable side effects of oral medications. Patients who elect to undergo neurosurgical procedures are typically amazed at the dramatic reduction in tone, but even after the stiffness is gone, their motor disorder and weakness remain. Eliminating spasticity does not cure cerebral palsy, and treating spasticity alone is not sufficient to optimize motor function in patients with cerebral palsy. Furthermore, although reduction in spasticity is desirable, it is not a prerequisite for improving strength and mobility in these patients.

STRENGTH AND PHYSICAL FITNESS

In addition to motor control, active, volitional movement requires strength and energy. The unbendable limbs of patients with cerebral palsy may feel "strong" to the practitioner or parent who tries to manipulate a patient's joints or stretch his or her stiff muscles. It can also be misleading when a nonambulatory patient who cannot sit is able to stand up on stiff legs, using his or her tone for support. Yet, patients with cerebral palsy fatigue more quickly and are less energy efficient than their "able-bodied" counterparts.²⁰ Patients with cerebral palsy may also have reduced lung function compared with other children their age.²¹ So, the child with cerebral palsy faces a daunting uphill battle struggling to use stiff, weak muscles in a growing body that moves slower and burns up more energy than normal. It is not surprising that these patients tend not to move.

Historically, children with cerebral palsy have not been expected to move very much. Not so many years ago, they were excluded from athletics and excused from physical education classes. Fortunately, things are changing. The merits of strength-training programs, once thought to be deleterious to patients with spasticity, are beginning to be recognized.^{22,23} Preliminary investigations indicate that aerobic exercise is at least as beneficial to these patients as traditional physical therapy²¹ and that exercise is an important factor in reduction of chronic pain associated with cerebral palsy.3 However, much more work needs to be done. For example, it is not known how much the inherent muscle properties, movement difficulties, weakness, or hypertonicity in cerebral palsy contributes to a patient's energy inefficiencies or whether increased oxygen consumption is more directly related to the deconditioning effects of a sedentary lifestyle.

GET THEM MOVING!

It is possible to dramatically increase the patients' energy level and increase ability and desire to move using a multifaceted, common-sense approach that centers on improving cardiovascular fitness. Regular aerobic exercise improves strength, physical fitness, and motor function.^{21,24,25} Insisting that patients participate in regular aerobic exercise also sends them the very powerful message that health and physical fitness are important and that having a disability need not imply poor health. There are a wide variety of adapted sports and other athletic activities in which these patients can participate (Table 4).

In addition to aerobic exercise, each patient needs an individualized home program that includes daily stretching, strengthening, weight-bearing, and balance activities. Goaldirected physical and occupational therapies are also important to help patients safely work toward maximal motor function, mobility, and independence. Therapies should be used only as an adjunct to a home program and not in place of one. Other promising new modalities include the use of neuromuscular electrical stimulation and isokinetic strength training to improve motor function in patients with cerebral palsy.^{22,26–28}

INDEPENDENCE

A person who is independent is able to make his or her own choices and pursue his or her own dreams in life. Most children with cerebral palsy will grow into adulthood and outlive their parents. Mobility is crucial to their health and well-being. Not only does mobility protect them from the numerous health consequences of a sedentary lifestyle, it also provides them with the opportunity to become independent. Mobility can mean the difference between growing up as an "observer" who watches the rest of the world go by or becoming a "participant" who is actively involved in the living, moving, and doing. Powered mobility (electric wheelchairs) and numerous assistive devices can further augment an individual's mobility tremendously, but equipment cannot completely substitute for strength and strategy.

Imagine what life would be like if you were completely dependent on someone else to take care of you. Imagine that you are so dependent that you are unable to support any weight on your arms or legs and you have to be lifted by a well-trained caretaker in order to transfer in and out of your power wheelchair. Even routine tasks require planning. You must ask for help to go to the bathroom and you need to schedule regular restroom breaks to ensure that you will have assistance when you need it. (I have patients

Table 4. Sports and Physical Fitness

Aerobics (water or land)	Martial arts*	
Aqua-jogging	Soccer	
Basketball*	Swimming	
Canoeing	Snow-skiing	
Dance*	T-ball	
Golf*	Therapeutic horsemanship*	
Ice-skating	Water skiing*	

*Most sports can be adapted for patients with cerebral palsy, such as "walker soccer" and sit-skiing (either water or snow). Water activities are accessible to nearly any individual, regardless of the severity of their motor disorder. The athletic activities listed here are only some of the sports in which individuals with disabilities can participate. The asterisk denotes sports that are exceptionally good balance activities. who restrict the amount of liquid they drink, sometimes severely, just so they do not have an accident in the middle of the day when their assistant is unavailable.) This degree of dependence severely limits ability to go out of the house, work, and socialize with friends unless one is lucky enough to have a personal assistant who is always available or one has very large, strong, understanding friends and coworkers.

Now imagine a different scenario: you still use a power wheelchair for locomotion, but you exercise to improve your endurance and gain enough strength in your legs to be able to stand briefly with assistance, pivot, and transfer. This degree of improvement in strength and ability will substantially reduce your risk for being dropped since you are no longer a full-lift transfer. It will reduce the risk of injury to your assistant and will make it possible for more people to assist you regardless of their size or strength. Furthermore, you are more likely to be able to use assistive devices such as a railing to make transfers even easier. Suddenly, even without being able to walk, you have more freedom and control over what you do and when you do it.

CONCLUSION

It is time to change the way that we in the medical profession look at patients with cerebral palsy. Although spasticity is one of the most tangible symptoms in patients with this group of heterogeneous disorders, these patients are more than just a composite of their spastic parts. They are real people with real goals, hopes, dreams, and feelings. Children with cerebral palsy start life wanting to succeed, despite the apparently overwhelming obstacles that many of them seem to face. Most children with cerebral palsy are capable of doing far more and achieving at much higher levels than the medical profession or society would have them believe. Managing the patient with cerebral palsy requires more than just reducing their spasticity or treating the complications that arise from living with a chronic motor disorder (eg, pain and fracture).

To make a difference in the lives of patients with cerebral palsy, we need to roll up our sleeves and get down in the trenches with them. We need to listen to them and try to see the world from their perspective. In order to listen to our patients, we must first be sure that they have a way to effectively communicate with us and with other people in their environment. We must teach them to set long-term goals for which they can strive, and then we must help them design a strategy and find the tools they need to tackle the hard work of reaching those goals. It is our responsibility to ensure that our patients' educational needs are addressed appropriately. It is our responsibility to teach our patients the importance of being healthy and physically fit, including the consequences of a sedentary lifestyle (first we must teach ourselves that having a disability does not preclude health and physical fitness). Doing our job right means that we become advocates for our patients, teaching society the importance of accessible environments and correcting society's misperception of what it means (and what it does not mean) to have cerebral palsy.

If, after reading this article, you have concluded that the comprehensive management of the patient with cerebral palsy is beyond the scope of most general child neurologists or that it is too difficult for one person to implement, then I would agree with you. Optimal management of a child with cerebral palsy requires a team of experts from multiple disciplines and a broad network of support systems within the community. Anyone can join the team at any time. Taking care of patients with cerebral palsy and doing it right requires a lot of hard work and commitment, but it need not be overwhelming. You do not have to do the work of the whole team (nor should you), and you do not need to know all of the answers in order to make a big impact. The only absolute requirement for team membership is that you listen to your patients and their families; they are the team leaders and are by far your greatest teachers.

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