A Pediatric PERSPECTIVE

Gillette Children's Specialty Healthcare

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A Range of Treatment Options Individualizes Spasticity Management

by Angela Sinner, D.O., pediatric rehabilitation medicine specialist and Debbie Song, M.D., pediatric neurosurgeon

In the United States, cerebral palsy occurs in approximately two children per 1,000, and as many as 1 million people are affected.[1] Cerebral palsy is a motor disorder affecting movement, posture and balance. It may be accompanied by disturbances of sensation, cognition, communication, perception, behavior and, potentially, seizures. The severity of those symptoms varies widely. However, 70 to 80 percent of patients who have cerebral palsy have spasticity.[2]

Spasticity is velocity-dependent resistance that affects muscle movement by increasing deep tendon reflexes and clonus. Spasticity is caused by a disruption in the baseline inhibitory signals to the stretch reflex, so muscles are stiff and movements are often jerky and awkward. Consequently, even the simplest activities of daily living—walking, using hands, talking, swallowing—may be affected. Additionally, the constant stress and abnormal growth forces associated with spasticity may cause muscle and joint contractures, as well as skeletal deformities. Successfully managing spasticity can increase function and quality of life, while reducing deformity. At Gillette Children's Specialty Healthcare, we provide a range of spasticity treatments to meet our patients' varied needs.

Spectrum of Spasticity Care

Our spasticity treatment decisions are based on a variety of factors, include the following: the patient's age, severity of spasticity and Gross Motor Function Classification System (GMFCS) level. We also consider the patient's goals, along with the family's access to follow-up care and ability to comply with the treatment recommendations. To meet patients' individual needs, we provide a range of treatments, and we may recommend combining treatments to effectively manage spasticity (see the case study on P. 3).

The spasticity management options Gillette offers include the following: therapy (physical, occupational and/or speech); bracing and splinting; oral or injected medications like botulinum toxin A; and surgical procedures such as implanting an intrathecal baclofen pump or performing a selective dorsal rhizotomy.

KEY INSIGHTS

- The severity of cerebral palsy symptoms varies widely, but most patients who have cerebral palsy will have spasticity.
- Oral medications to control spasticity will be chosen based on the individual's tone type, other symptoms and tolerance.
- Botulinum toxin A and phenol injections are well-suited for reducing focal tone issues or if the patient is too young for more generalized tone reduction treatments. They can also be used in combination with other spasticity treatments.
- Intrathecal baclofen therapy is used to provide a generalized reduction in tone. The implanted pump is refilled in clinic every few months, depending on the patient's medication requirements.
- Selective dorsal rhizotomy is a procedure that permanently reduces spasticity by cutting the nerve rootlets that transmit the abnormal signals associated with spasticity.

Inside

Some patients will require a combination of treatments to effectively manage their spasticity (see the case study on P. 3). To relieve spasticity, we may prescribe some or all of the treatments below.

Nonsurgical Treatments

Oral medications include benzodiazepines like diazepam (Valium) and muscle relaxants such as dantrolene (Dantrium) or baclofen (Lioresal). At times, dopaminergic drugs such as levodopa/carbidopa (Sinemet) and trihexyphenidyl (Artane) may be used to treat increased muscle tone. Oral medications are chosen based on the individual's tone type, other symptoms and tolerance. We begin with low doses and gradually increase them as needed. If a drug must be discontinued, we taper it off, avoiding the complications associated with rapid withdrawal.

Injectable medications like botulinum toxin A (Botox) decrease muscle spasticity, which can allow us to help improve range of motion and mobility. This medication is best for addressing focal tone issues or if the patient is too young for more generalized tone reduction treatments. Botulinum toxin A works at the neuromuscular junction to inhibit acetylcholine release. Once the drug has been administered into a muscle, the patient can more readily participate in physical or occupational therapy and be fitted with a brace or splint. Note: Botulinum toxin A is widely accepted as a treatment for children who have spasticity, but it is an off-label use for this medication.

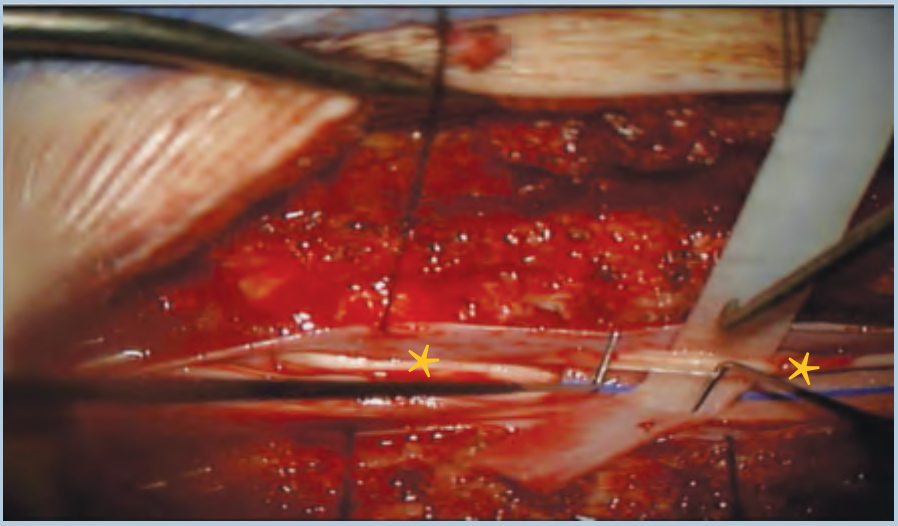
The location of the injection is based on a patient's needs and goals, and the dosage depends on the patient's size and prior exposure to the medication. At Gillette, our patients typically undergo conscious sedation before botulinum toxin A is injected. Botulinum toxin A has a temporary effect that usually lasts for three or more months.

Phenol neurolysis is another approach to reducing spasticity and tone. Phenol is dripped directly on the nerve to denature the nerve protein and thus damage the myelin sheath. As a result, the overactive signal is conducted more slowly through the nerve to the muscle.

Patients are usually sedated in the operating room to ensure their comfort while phenol is being administered. Sedation also helps them lie still, which allows for careful motor stimulation and localization of specific nerves. The effects of phenol typically last for approximately six months.

Neurosurgical Treatments

Prior to recommending a neurosurgical intervention, an interdisciplinary team (a neurosurgeon, a pediatric rehabilitation medicine specialist and an orthopedic surgeon) evaluates the patient. As part of the assessment, the patient undergoes a physical therapy evaluation and a gait and motion analysis. The team discusses a course of treatment with the family and may recommend a surgical intervention.



Close-up of a Selective Dorsal Rhizotomy

The edges of the spinal (dural) sac are retracted open with suture (black). Several nerve roots are exposed (yellow asterisks). A cotton pad is placed under one dorsal nerve root as it enters the spinal sac. Two hooks hold up a nerve rootlet that has been divided from the bigger nerve root. The hooks are used to electrically stimulate the rootlet. If stimulating the nerve rootlet results in an abnormal electrophysiological or clinical response in the muscles, we cut that nerve rootlet. Each nerve root is divided into five to 30 rootlets, and 20 to 40 percent of rootlets are cut.

Intrathecal baclofen therapy (ITB) is a drug delivery system that consists of a pump implanted in the abdomen and a catheter whose tip delivers liquid baclofen (Lioresal) directly to the intrathecal space (where the spinal cord, nerve roots and cerebrospinal fluid are contained). Because the medication is administered intrathecally, a much smaller dose is needed to reduce spasticity than is required when the oral medication is used. ITB therapy is well-suited for patients who require a generalized reduction in tone or whose goals include a focus on functional changes.

The therapy is also helpful for increasing patient comfort, reducing pain and making it easier to manage activities of daily living, such as dressing.

An external programmer is used to set the pump so it delivers the specified dosage of baclofen. The amount can be adjusted to achieve an optimal therapeutic effect. The pump is refilled with baclofen every few months in an outpatient setting. The timing of the refills depends upon a patient's dosage requirements, the concentration of medication used and the size of the pump implanted. Because the pump has limited battery life, it must be surgically explanted and replaced with a new pump after five to seven years.

Selective dorsal rhizotomy (SDR) surgery permanently reduces spasticity by interrupting abnormal signals being carried through the sensory or dorsal nerve roots to the spinal cord. The concept of SDR is to reduce high muscle tone by cutting portions of the nerve roots that carry abnormal signals.

During SDR, the lumbar nerve roots that transmit information to and from the muscles of the lower extremities are identified. Each individual sensory nerve root that carries information from the muscles to the spinal cord is then separated into five to 30 smaller threadlike rootlets, and each of the tiny rootlets is electrically stimulated. Rootlets that contribute to a child's increased muscle tone are identified by the abnormal electrophysiological and/or clinical response recorded in the muscles of the legs. The rootlets that demonstrate an abnormal response when stimulated are cut, while the rootlets that trigger normal responses when stimulated are not cut. Usually, 20 to 40 percent of the nerve rootlets are cut.

The procedure may be done for patients who have spastic diplegic cerebral palsy, are 4 to 10 years old, and are typically at GMFCS levels I, II or III. Usually, children assessed at those levels have impairments affecting their lower extremities, but can walk independently or with the help of a walker or crutches. For more information about patient selection for SDR, see Trost J P, Schwartz M H, Krach L E, Dunn M E, and Novacheck, T F. Comprehensive short-term outcome assessment of selective dorsal rhizotomy. *Developmental Medicine & Child Neurology* 2008, 50: 765–771.

Rehabilitation and Orthopedic Interventions Following SDR

After an SDR reduces spasticity and tone, patients require intensive rehabilitation so they can relearn muscle patterning. At Gillette , we recommend an extended inpatient rehabilitation stay. Children work on stretching, strengthening and movement. Therapy begins at a basic level: patients work on core strength and basic gross motor positions and movement, and their therapy is adjusted as they progress. Typically, patients continue intense outpatient rehabilitation therapy after discharge.

Approximately one to two years after a rhizotomy, our orthopedic surgeons reassess the child's gait and any orthopedic deformities. If required, our orthopedists may perform a single event multilevel surgery (SEMLS) to correct muscle contractures, bone deformities, weakness, poor motor control, impaired balance or other problems associated with cerebral palsy. SEMLS will also be followed by rehabilitation therapy.

Conclusion

Although most patients who have cerebral palsy also have spasticity, the severity of the spasticity varies widely, and there are a number of treatments to reduce spasticity and tone, enhance comfort and improve function. Having a range of spasticity management options allows us to individualize our care and match treatments with each patient's condition and goals.

[1, 2] *www.emedicinehealth.com/cerebral_palsy/article_em.htm, accessed 05/02/12*

Multiple Therapies Used to Manage Spasticity

A female patient has diffuse tone abnormalities as a result of schizencephaly. Her quadriplegic cerebral palsy is associated with significant spasticity and dystonia of the lower extremities, and she has mixed tone in her trunk. Consequently, taking care of her (bathing and dressing) is difficult. When she was 2 years old, she began receiving botulinum toxin A injections. When she was 4 years old, the decision was made to use a combination of treatments. She would have an intrathecal baclofen pump implanted to manage the spasticity in her lower extremities. Botulinum toxin A or phenol injections could also be used, as needed, to reduce the tone in her upper extremities.

During the baclofen pump implant procedure, the catheter tip was placed in her lower thoracic spine, and flow was established. The pump was pocketed on the right side in the abdominal area, and the catheter was tunneled between the pump and the catheter tip. The pump was filled with baclofen at 500 mcg/mL concentration. After a priming bolus, it was programmed to run at 100 mcg/day with simple continuous flow. The tip, catheter and pump were connected and secured. Both wounds were closed. The patient did not experience any complications postoperatively. The tone in her lower extremities was significantly improved. Periodically during the next few years, she also received botulinum toxin A injections to help reduce the spasticity in her trunk and extremities.

When she was 10 years old, a baclofen pump replacement was planned. Because the patient needed refills at 60-day intervals instead of the more customary 90-day intervals, the pump battery was depleted faster. She was implanted with a new larger pump (40 mL size), and it was connected to the existing catheter, which was still intact. Postoperatively, she had reduced spasticity in her lower extremities, so she could sit more comfortably in her wheelchair. She will continue to receive botulinum toxin A or phenol injections as needed to address the spasticity in her shoulder and torso.

Author PROFILES

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Pediatric Rehabilitation Medicine

Angela Sinner, D.O., specializes in pediatric rehabilitation medicine with a special interest in spina bifida, neurotrauma, and spasticity management. She received her doctor of osteopathic medicine degree from Des Moines University's College of Osteopathic Medicine in Des Moines, Iowa. She completed a physical medicine and rehabilitation residency at the University of Minnesota Medical School in Minneapolis, and then completed specialty training at Gillette Children's Specialty Healthcare through a fellowship in pediatric rehabilitation medicine. She has made numerous professional presentations on topics including pediatric concussion, posterior fossa syndrome, autonomic dysfunction in severe traumatic brain injury, and spinal cord injury evaluation. Her recent research has focused on intrathecal baclofen pump management as well as hypercalcemia incidence and treatment in spinal cord injury.

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Pediatric Neurosurgeon

Debbie Song, M.D., is a pediatric neurosurgeon who treats patients who have hydrocephalus, cerebral palsy, spina bifida, and brain and spinal cord anomalies. She has a special interest in Chiari malformations and spinal dysraphism. Song received her medical degree from the University of Michigan Medical Center in Ann Arbor, Mich. She also completed a neurosurgery residency, surgical internship and neurosurgery research fellowship there. Additionally, she completed a clinical research fellowship in the surgical neurology branch of the National Institute of Neurological Disorders and Stroke at the National Institutes of Health in Bethesda, Md. She finished a fellowship in pediatric neurosurgery at the University of Texas Southwestern Children's Medical Center in Dallas. She has extensive publications, including professional journal articles, book chapters and abstracts. She is a member of the Congress of Neurological Surgeons and American Association of Neurological Surgeons.

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Gillette Nationally Ranked in Pediatric Orthopedics and Pediatric Neurology/Neurosurgery

Gillette Children's Specialty Healthcare is now ranked 12th in the nation for pediatric orthopedics and 39th in the nation for pediatric neurology/neurosurgery in U.S. News Media Group's 2012-2013 Best Children's Hospitals rankings. Last year, Gillette ranked 25th and 44th in the two areas.

View the rankings and learn more at www.usnews.com/childrenshospitals

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