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. 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 these symptoms varies widely. However, 70 to 80 percent of patients who have cerebral palsy have spasticity. 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.

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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 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 skin or muscles (sensory root) is then separated into five to 30 smaller twinkly 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 electromyograph and/or clinical response recorded in the muscles of the leg. The rootlets that demonstrate an abnormal response when stimulated are cut, while the rootlets that trigger normal responses when stimulated are not cut. Usually, 10 to 40 percent of the nerve roots 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. Development Medicine & Child Neurology 2008; 50: 761-771.

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 to educate children on stretching, strengthening and movement. Therapy begins at a basic level; patients work on core strength and basic gross motor movements and posture, and their therapy is adjusted as they progress. Typically, patients continue intense outpatient rehabilitation therapy after discharge.

Approximately six to two years after a rhizotomy, an orthopedic surgeon reassesses the child’s gait and any orthopedic deformities. If required, our orthopedists may perform a single event multilevel surgery (SEMLS) to correct muscle contractions, 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 who have spastic diplegia receive successful results, patients with 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. Development Medicine & Child Neurology 2008; 50: 761-771.

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Conclusion

Although most patients who have cerebral palsy also have spasticity of the shoulder and torso, there are a number of treatments to reduce spasticity and tone, enhance comfort and improve function. Having a range of spasticity management options allows families and therapists to choose the care and match treatments with each patient’s condition and goals.

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, the main core of her left hand (striking 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 from 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 normal functioning. The pump and catheter 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 30-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 shoulders and torso.
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 to six months.

Pharmacology

Botulinum toxin A is a neuromuscular inhibitor. It works by inhibiting 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 prescribed for other indications, such as managing spasticity or improving 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.

Oral medications include benzodiazepines like diazepam (Valium) and muscle relaxants such as dantrolene. Oral medications are chosen based on the individual’s needs, other symptoms and tolerance. We begin with low doses and gradually increase them as needed. If a muscle drug is discontinuing, 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.

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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.

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Rehabilitation and Orthopedic Interventions Following SDR

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Conclusion

Although most patients who have cerebral palsy also have spasticity, the severity of the spasticity varies widely. Gillette has a number of treatments to reduce spasticity and tone, enhance comfort and improve function. Having a range of spasticity management options allows our rehabilitation and orthopedic core teams and match treatments with each patient’s condition and goals.

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**Spectrum of Spasticity Care**

Our spasticity treatment decisions are based on a variety of factors, including 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. 5).

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.

**Spinal Cord Injury**

Spinal cord injury (SCI) affects the body’s ability to control muscle movement. SCI is the most common cause of catheter-blocked bladder. The spasticity management options Gillette offers 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. 5).

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**Epilepsy**

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