

Dear Physicians, Parents, and Caregivers:

On behalf of the Child Neurology Society and the Child Neurology Foundation, it is our pleasure to team with Allergan, Inc. to introduce you to a new publication that covers the current spectrum of treatment options for spasticity. It is our hope that you will find this an important tool in determining the highest quality care for your child.

Throughout North America, child neurologists provide nearly eighty percent of direct and consultative care for children and youth with disabilities and serious health conditions. This pamphlet serves as a valuable tool and underscores our profession's commitment to that care.

We want to thank Dr. Michael Noetzel and his team for preparing a useful and easy-to-understand publication, which will be available in the offices of child neurologists throughout the U.S. and Canada, as well as by request from the Child Neurology Foundation at 877-645-4319 or by email at [cnf@childneurologyfoundation.org](mailto:cnf@childneurologyfoundation.org).

The pamphlet will also be available on the Foundation's website at [www.childneurologyfoundation.org](http://www.childneurologyfoundation.org) where you may simply print a copy. We are honored to be associated with this project and to continue to work in partnership with parents and caregivers who are our patients' strongest advocates.

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current treatment options for  
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# common approaches to the management of spasticity

## introduction

Spasticity is a common feature of many chronic motor disorders affecting infants, children and adolescents. A widely used definition of spasticity is that it is an exaggerated response to passive movement of a limb in which there is velocity dependent resistance of a muscle to stretch. Spasticity is characterized by excessive and inappropriately timed activation of skeletal muscles, which may interfere with a child's ability to move voluntarily in a normal fashion. However, spasticity also affects passive movement and posture. In a recent National Institutes of Health workshop, it was emphasized that spasticity must be differentiated from other manifestations of impaired movement, especially dystonia and rigidity, since treatment decisions are based upon the type of abnormal tone. In the pediatric population spasticity is most commonly found in children with cerebral palsy. However, spasticity can result from damage or abnormal development of nerve cells or pathways controlling motor movement either in the brain or spinal cord. Thus, spasticity is not uncommonly seen in children with traumatic and hypoxic injury to the brain, stroke, spinal cord injury and brain tumors.

Spasticity may interfere with a child's control of voluntary movement, coordination, exercise tolerance and range of motion in the joints. As a result, spasticity can limit activities of daily living and may cause pain and disturbed sleep. In more severely affected individuals, patient care is often more difficult. Over time spasticity is associated with reduced protein synthesis and thus impaired muscle growth. If untreated there may be permanent shortening of muscles (contractures) and the development of bone and joint deformities.

## evaluation and goals of spasticity management

Evaluation of a child with spasticity includes a thorough neurological examination to document the pattern and severity of spastic involvement and to determine how the muscle hypertonia is interfering with function and/or patient care. The exam should include standardized measures of tone, as well as assessment of motor performance and functional ability. In selective cases, gait analysis may be helpful. Treatment of spasticity is designed not merely to decrease the spastic hypertonia, but ultimately to improve a child's functional capabilities and, thus, promote their quality of life and hopefully their greater independence in society. Spasticity management can minimize pain, as well as decrease the incidence of muscle contractures. Again, for the more severely involved patient, spasticity management may be designed to facilitate hygiene, to reduce the requirements of care giving and to minimize orthopedic complications such as hip dislocation.

### REHABILITATION THERAPY

Treatment carried out by physical and occupational therapists is a mainstay in the care of children with disorders causing spasticity. Numerous therapy techniques may have a role in the management of spasticity ranging from passive range of motion to "stretch out" muscles to inhibitory casting, assisted ambulation, and strategies for daily living skills. Strengthening of spastic muscles may play a critical role since weakness may be a greater cause of disability than spasticity. To promote muscle relaxation, tone-inhibiting positioning techniques can be utilized. The proper seating of patients produces optimal postural alignment and thus improves the distribution of weight-bearing forces. Appropriate bracing and orthotics are a useful component of many therapy regimens. Aqua therapy and the application of heat, cold and electrical stimulation, as well as other physical modalities administered by therapists, have been used effectively as adjuncts in decreasing spasticity. In addition, any other treatment modality used to manage spasticity is most likely to benefit those patients receiving ongoing rehabilitation therapy.

### PHARMACOLOGIC MANAGEMENT

Medication has been a major component in the treatment of spasticity in adults over the last several decades. However, few appropriately designed studies have examined the role of oral anti-spasticity medication in improving a child's functional capability. Commonly employed medications treating spasticity in the pediatric population include diazepam, baclofen, tizanidine, and dantrolene. The mechanism of action of the first three agents appears to be through altering the excitatory and inhibitory neurotransmitters in the spinal cord, while dantrolene acts directly to inhibit the contraction of skeletal muscles. The side effects of dantrolene, which include weakness, fatigue, diarrhea and potential liver injury, can limit the effectiveness and duration of treatment. Additionally, the benefit of diazepam, baclofen and tizanidine may be overshadowed by side effects such as drowsiness, confusion, imbalance and excessive low tone. In view of these considerations, utilization of any oral anti-spasticity medication clearly must be justified by ongoing demonstrable benefit to the individual patient.

## INTRATHECAL BACLOFEN THERAPY (ITB)

In patients with more severe or extensive spasticity that is interfering with quality of life, intrathecal baclofen therapy can be employed to reduce tone. The components of an ITB system include: a pump implanted under the skin on the abdomen which infuses the drug at a pre-determined rate; a catheter which delivers the drug to the intrathecal space of the spinal cord; and a programmer which allows for adjustable and precise dosing. ITB therapy should be considered in pediatric patients whose severe spasticity interferes with function or care and those who have painful spasticity-related spasms. In addition, children and adolescents who are refractory to other treatments or unable to tolerate side effects from oral medications are also reasonable candidates. ITB formerly was utilized mainly in patients with predominantly lower extremity spasticity; more recent emphasis has been on those patients with quadriplegic cerebral palsy and mixed cerebral palsy with dystonia. Relative contraindications to ITB therapy include: spasticity affecting predominantly the arms; exceedingly poor trunk control; and families who cannot fully commit to the requirements of ITB therapy maintenance (pump refills every 2–6 months). In addition to device and procedure complications of ITB therapy, adverse drug related events have been documented. Even though the dose of intrathecal baclofen is lower than oral medication, side effects such as somnolence and hypotonia have been reported. Signs of overdose range from light-headedness and excessive weakness and low tone to respiratory depression, seizures and loss of consciousness. Withdrawal from the medication is a potential medical emergency, presenting as itching, altered mental status, exaggerated rebound spasticity and muscle rigidity that may advance to rhabdomyolysis and organ failure.

## BOTULINUM TOXIN

Intramuscular injection of botulinum toxin reduces spasticity by inhibiting the release of the neurotransmitter (acetylcholine) responsible for transforming nerve impulses into muscle contraction. Because the effects of botulinum injections are local, dosed dependent, and temporary, this treatment is best utilized in those patients who have specific "problem muscles" that interfere with major functional aspects of living such as ambulation or upper extremity movement. A clinically apparent affect is most notable 2–4 weeks after injection and ongoing benefit may last up to 3 to 6 months. Thus, serial injections often are required. In smaller muscles (such as those in the forearm and hand), EMG guidance of the injections may be required. Most children tolerate the series of injections with only local anesthetic, but in some cases general anesthesia may be necessary. Botulinum injections can be employed with other therapies including intrathecal baclofen and dorsal rhizotomy (see below). Systemic side effects are rare, but pain on injection and a transient flu-like syndrome have been noted. A variety of studies, mainly on children with cerebral palsy, has documented reduction in spasticity and functional improvement in the injected muscle groups.

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## ORTHOPEDIC SURGERY

Even with optimal treatment, over time spasticity may progress to cause muscle contractures and bony deformities. Thus, children with spasticity may eventually require orthopedic surgical intervention. The most common orthopedic procedures carried out in this population include tendon releases for fixed contractures at the ankle, surgery to correct hip subluxation or dislocation and instrumentation of the spine for scoliosis. The benefit of orthopedic surgery is most pronounced in patients who have achieved some degree of skeletal maturation (typically between ages 6 and 12). A single multi-level surgical intervention (if feasible) is most advantageous, as opposed to multiple procedures each with their individual post-surgical recovery time. All of these surgical interventions typically will mandate intensification of a physical therapy program to strengthen and improve the range of motion in the surgically improved extremity.

## SELECTIVE DORSAL RHIZOTOMY (SDR)

SDR is a neurosurgical technique carried out on the lower spinal cord designed to reduce moderate to severe spasticity in the legs. Bone covering the first two lumbar segments of the spine is removed. Individual sensory nerve roots are identified and those that demonstrate excessive activity as determined by electromyography are severed. Post-operatively the child is enrolled in an intensive program of physical therapy. Clinical studies have demonstrated that the combination of SDR and physical therapy is beneficial in reducing spasticity in children with spastic diplegic cerebral palsy. A positive, but less, robust effect on gross motor function also has been observed, with a direct relationship between the percentage of dorsal nerve roots transected and functional improvement. In view of the surgical approach in SDR, concern has been raised about the long-term risk of spinal deformity and an adverse effect on bladder function. Most studies, however, have suggested that these risks are minimal when the surgery is carried out using the limited two segment laminectomy. Persistent loss of strength following SDR also has not been objectively demonstrated.

Patients most suitable for SDR are those with spastic diplegia or mild quadriplegia who have some degree of ambulatory capability, but whose full potential is limited by lower extremity spasticity. Most of these individuals have a history of prematurity or evidence of periventricular leukomalacia on imaging studies. Surgery typically is carried out on children between the ages of 4 and 8 years. Unsuitable candidates for SDR include those whose lower extremity spasticity is a result of congenital or neonatal central nervous system infection and head trauma or hypoxic brain injury after the newborn period. In addition, children with very weak or low tone abdominal muscles and those who have predominant dystonia or ataxia, as well as those with limited potential for functional gains, are less likely to benefit from a SDR.

**conclusion** Treatment of spasticity remains a major therapeutic challenge in children with chronic motor disorders. Families often have a great deal of uncertainty about what treatment approach may be best for their child. In many ways the well-trained pediatric neurologist is an ideal physician to assist the family in making appropriate decisions regarding which spasticity intervention is most likely to produce benefit for their child.