

CME Article

Spasticity Management in Children

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Target Audience

This CME activity is intended for physicians, medical students and nurse practitioners. Pediatric emergency department physicians, emergency physicians, pediatricians and family practitioners will find this information especially useful.

Learning Objectives

After completion of this article, the reader will be able to:

1. Identify the clinical manifestations of spasticity.
2. Understand the importance of early comprehensive treatment.
3. Compare the benefits and potential complications of focal, regional and systemic treatments.

Spasticity is the hallmark of an upper motor neuron disorder. It is commonly observed as a result of damage to descending motor pathways at cortical, brainstem or spinal cord levels. The etiologies include, but are not limited to: traumatic brain or spinal cord injury, cerebral palsy, stroke, anoxia, infection and collagen vascular disease.

Spasticity has been defined as a velocity-dependent increase in resistance to stretch.¹⁻³ It is also commonly associated with hypertonicity, hyperreflexia, clonus and spread of contraction beyond the muscles stimulated.²

The earliest clinical manifestations of spasticity are in the form of hip extension, opisthotonus and cortical thumbing. In children with spasticity, abnormal movement patterns can also be recognized as components of persistent primitive reflex patterns such as the asymmetric tonic neck reflex, symmetric tonic reflex and the tonic labyrinthine reflex.⁴ These are some of the earliest markers of abnormal neurologic maturation. The abnormal movement patterns can be seen in limbs as the child attempts voluntary movement, triggered by passive positioning, in response to sensory stimuli, or as an "overflow" of uninvolved limbs.

Patient evaluation should include identifying when spasticity is interfering with move-

Abstract

Spasticity, or an abnormal increase in muscle tone, is a cause of significant impairment in children with central nervous system disorders. Identifying the clinical manifestations and functional implications of spasticity in the first few years of life will encourage appropriate growth and development in the affected child. Early and comprehensive treatment of spasticity will prevent complications such as muscle contractures and bony malalignment. These sequelae can manifest into more severe medical complications such as osteomyelitis from pressure ulceration or respiratory compromise resulting from scoliosis. This article will focus on the benefits, side effects and potential complications of focal, regional and systemic treatments for spasticity in children.

ment and when the patient is relying on spasticity for movement.

In other words, does the spasticity interfere with function such as gait, activities of daily living skills (ADL) or cause discomfort or difficulty with caregiving? Lastly, the unrelenting increase in abnor-

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mal muscle tone can progress to a musculoskeletal deformity. For example, hip adductor and flexor spasticity can create pelvic obliquity causing a scoliosis. The scoliosis can eventually progress to respiratory compromise.

The goals of treatment are two-fold. This should include functional and “technical” improvement. Functional goals encompass improvements in gait pattern, ease of caregiving, pain relief, decrease spasm frequency and increased ADL skills. The technical improvements are typically an increase in passive range of motion (using a goniometer) and a decrease in spasticity (Modified Ashworth Scale).

Initial Patient Management to Reduce Spasticity

A major critical factor in the initial patient management is the prompt treatment and avoidance of noxious stimuli. Several medical complications can exacerbate spasticity in patients with an upper motor neuron disorder. These include, but are not limited to: urinary tract infections, bladder calculi, fecal impaction, bladder distension, pressure sores, muscle contractures, deep vein thrombosis, tracheitis, upper respiratory tract infection, pneumonia, gastroesophageal reflux and dysphagia.^{1,3,4} Symptoms from these illnesses will increase a patient’s spasticity and, if treated immediately, will assist in the successful reduction of spasticity.

A second important factor in the initial patient management of spasticity is related to therapeutic intervention. These would include positioning, exercise, formal therapies (PT, OT SLP) splinting or casting and modalities. Proper sitting position or bed positioning is important in reducing spasticity. Midline and neutral joint positioning is crucial in maintaining proper muscular balance. Frequent stretching will also improve joint range of motion (ROM). Occupational, physical and speech therapies are essential to improve strength, function and in prevention of abnormal motor patterns. In addition, certain modalities can be attempted

to reduce spastic muscles such as heat, cold, neuromuscular electrical stimulation or therapeutic electrical stimulation. Lastly, orthoses (splints) and serial casting can assist in the improvement in ROM of a joint and in decreasing reflex tone by positioning the limb on a tonic stretch.

Systematic Spasticity and Use of Pharmacologic Intervention

When spasticity is generalized (involving the axial and extremity muscles), consideration should be given to the use of systemic medications. Typically in children, Lioresal (Baclofen), Tizanidine (Zanaflex), Diazepam (Valium) and Dantrolene (Dantrium sodium) are used in an attempt to reduce spasticity.^{1,3}

Consideration should be given to the drug’s site of action when recommending a specific medication. For instance, Lioresal’s (Baclofen) sites of action are the GABA ‘B’ receptors in the spinal cord. Therefore, it is usually the drug of choice for spinal cord injury and multiple sclerosis. The site of action for Dantrolene (Dantrium) is at the level of the intrafusal and extrafusal muscle fibers. Consequently, it is recommended for use in cerebral causes of spasticity. Tizanidine’s (Zanaflex) site of action is the alpha-adrenergic receptors in the spinal cord and supraspinally. It is the drug of choice in spinal cord injury, multiple sclerosis and stroke. Diazepam’s (Valium) sites of action are the benzodiazepine sites in the brainstem reticular formation and spinal cord. Therefore, it is useful in spinal cord injury. All of the systemic medications have sedation as a potential side effect. Other important considerations should be given to the precautions, side effects, dosing and mode of action when determining the most beneficial medication for the patient.

The advantages of systemic medications are that they are non-invasive and are not permanent. The medication can be weaned off gradually in order to prevent irritability and side effects since these are cen-

trally-acting agents. These drugs can be very effective management for some patients.

The disadvantages include difficulty in achieving a steady state, side effects of sedation, muscle weakness and hypotonia, physical tolerance, and for some, difficulty in following a schedule.

Intramuscular Pharmacologic Treatment of Spasticity

The use of neurolytic blocks can be effective management when a limited number of muscles need spasticity reduction. These treatments include botulinum toxin type A (Botox), botulinum type B (Myobloc) and motor point blocks with phenol.

Botulinum toxin type A is used more readily in pediatrics due to a much lower protein load.^{3,6,9} than type B. The toxin is produced by the anaerobic bacterium *Clostridium botulinum*. It targets the presynaptic acetylcholine neuromuscular junction thereby preventing the release of acetylcholine. The onset of action is between 12 to 72 hours, and the duration of effect is approximately four months.⁶ The maximum total dose per visit is 12units/kg up to 400 units.^{5,6} Re-injection should not occur sooner than every three months due to the risk of antibody formation and subsequent resistance to the drug.

The selection of patients depends on three basic principles. There should be the presence of dynamic contractures, the lack of progress with non-invasive approaches and the ability to target specific muscles or muscle groups.

The advantages of the drug are that it is not a permanent treatment, the effects are localized and there is evidence to support the efficacy in reducing spasticity and improving function.⁷ The disadvantages include the fact that the patient may need repeat injections, the drug is more expensive than other injectable neurolytics and the patient may develop antibodies to the medication. The contraindications are virtually non-existent. Fever rarely occurs, too high a dose may cause

excessive weakness and there may be local irritation from the injection itself.

A second neurolytic block used in pediatrics are motor point blocks with phenol.^{8,9} Phenol is a drug that temporarily ablates the neuromuscular interaction. It is a caustic and irreversible neurotoxic agent. The duration of effect is approximately seven months. Motor point localization is accomplished using an electrical pulse generator, therefore there is the need to place the child under anesthesia. The drawbacks of the procedure are that it is a painful medication and procedure, it requires general anesthesia during administration and there is the potential for painful sensory dysesthesias.

Neurosurgical Treatment for Spasticity

When spasticity is no longer manageable by non-invasive methods, then consideration should be given to two alternatives, Intrathecal Baclofen (ITB) Pump Implantation or Selective Dorsal Rhizotomy.

Long-term delivery of baclofen directly to the intrathecal space is an effective treatment for patients with intractable spasticity or dystonia. It uses an infusion system consisting of an implantable pump, catheter and external programmer. Baclofen delivered intrathecally allows higher CSF concentration than oral baclofen therapy with lower side effect risks.^{1,10-13} The efficacy depends on the Screening Trial Bolus injection administered via lumbar puncture. The onset of action of intrathecal baclofen is 30-60 minutes and the peak effect is in four hours.

The potential candidates are those with intractable spasticity or dystonia and systemic medication failure. Children over the age of 4 or of sufficient body mass to support an implantable pump are usually selected for ITB.^{11,12}

The child should have underlying weakness, which is not amenable to Selective Dorsal Rhizotomy. The spasticity management team and the family should have clear and realistic goals for the patient considered for

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implantation. The family/parent should be compliant with the recommended treatment as exhibited by their involvement with previous and current spasticity management. Lastly, there should be an absolute improvement in spasticity following the ITB screening trial dose.

One advantage of ITB is that it is a reversible procedure. The pump can be removed from the patient. A second advantage is that the intrathecal baclofen dose is titratable by easily utilizing the external programmer.

There are a few disadvantages to ITB pump implantation. The pump is a mechanical system run by a battery that will need to be surgically replaced approximately every four years. Statistically, there is approximately a 10% risk of infection and a 20% risk of catheter problems.¹¹ ITB pumps require ongoing maintenance in order to refill the medication and to program the pump for an appropriate therapeutic effect.

The explicit contraindications include infection at the time of screening or implantation or a history of hypersensitivity (allergy) to oral baclofen.²

A second very effective neurosurgical treatment for intractable spasticity is Selective Dorsal Rhizotomy. This is a procedure where there is a neurosurgical ablation of a select proportion of dorsal (sensory) nerve rootlets. Intraoperative electromyographic monitoring is used to assist with root identification and appropriate sectioning in the operating room. Surface electrode placement is over a variable number of target muscles with L2-S2 innervation (to include the contralateral limb, an upper extremity and the face). The roots or divided rootlets are then stimulated and evidence of a sustained response in multiple segments of the ipsilateral leg and spread of response to the contralateral leg, face or arms will be sectioned.

Patients selected should have pure spasticity as their movement disorder. There should be absent athetosis, ataxia, dystonia and rigidity.^{3,4,11} Function should be limited primarily by spasticity and not being used for functional benefit. For example, spasticity in the lower

limbs of patients with spinal cord injury sometimes enables them to do a stand-pivot transfer utilizing limbs that are “stiff”. There should be absence of profound underlying weakness and adequate trunk control.¹⁴

The patient should have good selective motor control and righting responses with some degree of forward locomotion.¹⁴⁻¹⁶ Young patients between the ages of 3-8 are usually preferable because motor patterns are usually “fixed” after that age.^{3,4,16} The patient should have minimal or no joint contractures or spinal deformity.

Lastly, the patient should have adequate cognitive ability to follow directions and participate in therapy. The family should be supportive, interactive and committed to postoperative therapy. This consists of six to nine months of intensive therapy if improved function is the goal.

The potential adverse effects include hypotonia, loss of sensation, hip subluxation/dislocation, unmasked underlying muscular weakness, bladder dysfunction and spinal deformity.

The advantage of SDR is that it is a one-time, permanent procedure used to dramatically reduce spasticity. There is evidence in the literature for efficacy in reducing spasticity and improving function in children with spastic diplegic cerebral palsy.¹⁴

One disadvantage is that since it is a permanent procedure, the patient may still need some of the spasticity to function. A second disadvantage is that it is not effective for treating dystonia.

Orthopedic Surgical Intervention

There is an important role for the judicious use of orthopedic intervention in the overall plan for rehabilitation. When there is static deformity (muscle contracture or bony deformity), intervention is critical for improving patient function. The spasticity management team must analyze the effects of growth on the problem, with and without treatment. The purpose and expectations for

surgery must be clear to all involved in the care of the child.

There are several orthopedic surgical approaches used in patients with spasticity-related dysfunction. These include: lengthening muscles with spasticity to improve function, transferring tendons to redirect muscle forces, releasing muscles and joint capsules with contractures and bone reconstruction for bony deformity.

The combination of spasticity reduction and orthopedic correction of bony deformity can probably maintain muscle length throughout the years of growth and minimize surgical lengthening of muscles.

Conclusion

In children with neurological injury, spasticity is a common denominator necessitating comprehensive treatment to reduce discomfort and improve functional outcome.

Spasticity causes loss of selective motor control and balance deficits. The inability to stretch these muscles through normal play results in fixed muscle contracture and abnormal skeletal forces ultimately leading to bony deformity. If spasticity is reduced in the growing child, many of the secondary complications can be handled by conservative, non-surgical means.

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QUESTIONS

- 1. Etiologies of spasticity may include all of the following except:**
 - a. Anoxic encephalopathy
 - b. Subarachnoid hemorrhage
 - c. Cauda equina syndrome
 - d. Bacterial meningitis

- 2. One of the earliest signs of spasticity in infants is:**
 - a. Asymmetric tonic neck reflex
 - b. Hip extensor thrust
 - c. Ankle clonus
 - d. Hyperreflexia

- 3. Which of the following medical illnesses can exacerbate spasticity?**
 - a. Urinary tract infection
 - b. Gastroesophageal reflux
 - c. Fecal impaction
 - d. All of the above

- 4. All of the following are important considerations for the use of neurolytic blocks to treat spasticity except:**
 - a. The presence of dynamic muscle contracture
 - b. Systemic spasticity involvement
 - c. Lack of progress with non-invasive approaches
 - d. Ability to target specific muscles or muscle groups

- 5. Which of the following treatments for spasticity is NOT beneficial for choreoathetoid cerebral palsy?**
 - a. Selective Dorsal Rhizotomy
 - b. Intrathecal Baclofen Pump
 - c. Botulinum toxin type A
 - d. Phenol motor point blocks

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