Spasticity

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Course Description
Spasticity can be a significant problem in upper motor neuron disorders. Consequences of spasticity can include the development of contractures or chronic pain, functional problems with self-care and mobility, seating problems, difficulty sleeping, as well as cosmetic appearance problems. A good understanding of the pathophysiology of spasticity is essential for proper management of this problem. This course will aid the physician understand this pathophysiology. Furthermore this course gives a guide to the evaluation and comprehensive management of spasticity in both pediatric age groups as well as adults.

Intended Audience
This course is intended for Neurologists, Physiatrists, and others who practice neuromuscular, musculoskeletal, and electrodiagnostic medicine with the intent to improve the quality of medical care to patients with muscle and nerve disorders.

Learning Objectives
Upon conclusion of this program, participants should be able to:
(1) identify pathophysiology of spasticity.
(2) perform an evaluation of an individual with spasticity.
(3) identify different treatments available for spasticity management.
(4) summarize indications and pros and cons of different spasticity treatments.
(5) improve diagnosis of spasticity and management of patients with spasticity.

Activity Profile
This enduring material activity is a reproduction of the printed materials from a course at the AANEM Annual Meeting (October 6-9, 2010). Physician participation in this activity consists of reading the manuscript(s) in the book and completing the clinical and CME questions.

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Duration/Completion Time: 2 hours

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Spasticity

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Dr. Sheean is on the Advisory Board for Ipsen. Any conflict of interest was resolved according to ACCME Standards.

Dr. McGuire is on the Allergan and Medtronix Speakers Bureaus. Any conflict of interest was resolved according to ACCME Standards.

All other authors/faculty have nothing to disclose.

Course Chair: Stephen Kishner, MD

The ideas and opinions expressed in this publication are solely those of the specific authors and do not necessarily represent those of the AANEM.
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Director  
EMG and Neuromuscular Services  
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Dr. Sheean is the director of electromyography (EMG) and neuromuscular services at the University of California, San Diego. He graduated from medical school, internal medicine residency, and neurology residency in Australia. He underwent further training and performed research in clinical neurophysiology at The National Hospital for Neurology and Neurosurgery, Queen Square, London. Following that, he joined the staff at Queen Square and St. Mary’s Hospital, Paddington, as a consultant clinical neurophysiologist. While at Queen Square, he developed an interest in botulinum toxin treatment, particularly for task-specific focal hand dystonia and spasticity, and in the pathophysiology of spasticity. He later moved to the University of California, San Diego.
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The Pathophysiology of Spasticity

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Clinical Professor of Neurosciences
Director, Neuromuscular Division
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San Diego, California

INTRODUCTION

Following a lesion of the upper motor neurons, several types of muscle overactivity can develop. Spasticity is a well-known type of this muscle overactivity and is a form of hypertonia that is velocity-dependent and length-dependent and is due to hyperexcitability of tonic stretch reflexes. However, there are several other forms of muscle or motor overactivity that can be seen (see Table 1). Most of these are thought to arise as a result of disinhibition of various spinal reflexes caused by interruption of the upper motor neuron pathways. However, the mechanisms underlying one form of motor overactivity—spastic dystonia—are unclear. It is important to understand that these types of motor overactivity, although often seen together in various combinations, are caused by separate and distinct spinal mechanisms and are not just some variations of spasticity.

SPINAL REFLEXES AND SUPRASPINAL CONTROL

Spinal reflexes are under supraspinal control by descending motor pathways that are predominantly inhibitory or excitatory. In humans, it appears that the corticospinal or pyramidal tract contributes very little to the upper motor neuron syndrome. Other upper motor neuron pathways (parapyramidal) are more important.

The main inhibitory pathway is the dorsal reticulospinal tract, which ascends in the spinal cord adjacent to the corticospinal tract. The main excitatory pathways are the ventral reticulospinal tract and the vestibulospinal tract, which are located elsewhere in the spinal cord and separate from each other. All these pathways arise in the brainstem but only the dorsal reticulospinal tract receives input from the cortex, which is excitatory. These anatomical and functional differences allow for different clinical patterns determined by the location and extent of the lesion. Whether the hyperexcitability of spinal reflexes is simply due to a shift of physiological balance to net disinhibition, the way that heart rate is determined by a balance between sympathetic and parasympathetic drive, is unknown. Neural plasticity changes, such as collateral sprouting or denervation hypersensitivity, also have been proposed. Recently, down regulation of a potassium-chloride cotransporter (KCC2) has been found in the rat model of spinal cord injury. This leads to impaired postsynaptic inhibition and spasticity and can be reversed by brain-derived neurotrophic factor (BDNF).

<table>
<thead>
<tr>
<th>Table 1 Types of motor overactivity in upper motor neuron syndrome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hyperkinetic</td>
</tr>
<tr>
<td>(involuntary movements)</td>
</tr>
<tr>
<td>Spasms (flexor, extensor, adductor)</td>
</tr>
<tr>
<td>Great toe extension</td>
</tr>
<tr>
<td>Associated reactions</td>
</tr>
<tr>
<td>Mass movements</td>
</tr>
</tbody>
</table>

* Includes active and passive movement
MOTOR NEURON EXCITABILITY

While net disinhibition of spinal reflex pathways appears to play a central role in the development of these motor overactivities, other mechanisms have been postulated. For example, muscle spindle hypersensitivity as a result of increased gamma efferent drive was suggested as a possible mechanism of increased static (tendon reflexes) and tonic (spasticity) stretch reflexes. However, evidence of this has not yet been found. Another possibility is increased intrinsic excitability of the motor neurons and evidence of this has been found in cat models of spasticity and in humans with spasticity. The mechanisms underlying this intrinsic hyperexcitability is an increase in persistent inward currents (PIC), which are strong determinants of the level excitability of a motor neuron and are under the control of descending serotonergic and noradrenergic pathways. Following an acute spinal lesion, persistent inward currents decrease dramatically leading to hypoexcitability of the motor neurons and a loss of reflex activity. This corresponds with the period known as spinal shock. Later, persistent inward currents recover and become increased above baseline, producing plateau potentials leading to spontaneous and exaggerated flexor spasms.

Many spinal reflex pathways have been studied electrophysiologically in spasticity and many have been found to be abnormal. However, it has been difficult to correlate any single spinal reflex abnormality with the presence and severity of spasticity. Currently, reduction of reciprocal 1a inhibition and of autogenetic 1b inhibition are strong candidates.

CLINICAL EVALUATION

One difficulty studying spasticity clinically and electrophysiologically is determining whether spasticity is actually present. In an upper motor neuron lesion, hypertonia is often produced by a combination of spasticity and stiffness of soft tissues. Thus, in some studies, hyperreflexia of tonic stretch reflexes has been absent in patients thought to have spasticity clinically. Recognizing the difference in hypertonia due to spasticity and hypertonia due to soft tissue stiffness is difficult but important for therapeutic reasons. Hypertonia due to soft tissue stiffness will not respond to antispasticity medications or chemodenervation. The often used clinical scales for evaluating spasticity is the Ashworth or Modified Ashworth scale, which cannot distinguish spasticity from soft tissue hypertonia. The Tardieu scale seems to do this better because it relies on the velocity dependence of spasticity. Needle electromyography (EMG) and examination under anesthesia can be helpful.

DISORDER OF MOVEMENT

Because spasticity, by definition, is a passive entity, evaluated with the subject at rest, its relevance to movement has been questioned. Studies have shown that tonic stretch reflexes in actively contracting muscles are not exaggerated in patients with spasticity. However, natural reflex modulation during functional movements is impaired, which could interfere with movement.

Patients with upper motor neuron lesions often exhibit abnormal cocontraction of agonist and antagonist muscle pairs during active movement, which clearly interferes with movement. For example, active extension of the elbow is impaired by simultaneous cocontraction of elbow flexors. It could be argued that the action of the elbow extensors produces a passive stretch of the elbow flexors, eliciting a tonic stretch reflex in the elbow flexors, similar to spasticity. However, isometric studies where stretch of the elbow flexors did not occur show cocontraction. In other studies, contraction of the antagonist muscle (e.g., elbow flexors) occurred before contraction of the agonist muscles (e.g., elbow extensors) had begun. In some cases, the antagonist contraction overwhelms the agonist contraction producing the opposite movement. Patients attempting to extend their fingers to open the hand often experience increased flexion of the fingers. Attempted dorsiflexion of the ankle may result in plantarflexion. This indicates that the upper motor neuron signal for contraction is not confined to the agonist muscles alone, probably due to failure of reciprocal inhibition, most likely at the level of the spinal cord.

While motor overactivity, which is considered a positive phenomenon in the upper motor neuron syndrome, can interfere with active movement, it is the negative phenomena, such as weakness, incoordination, and fatigue, which produced most impairment of movement. Spasticity and spastic hypertonia can interfere with passive movement, as can soft tissue stiffness, joint calcification, and contracture. Joint calcification and contracture limit the range of movement, whereas soft tissue stiffness increases the resistance to movement (i.e., tone) and possibly also the range of movement. Contracture can exist without hypertonia. The neurological consequences of an upper motor neuron lesion and the biomechanical complications are not separate (see Figure).

![Figure](https://example.com/figure.png)

*Figure* The neurological consequences of an upper motor neuron lesion and the biomechanical complications are not separate.

UMN = upper motor neuron lesion
OTHER TYPES OF MOTOR OVERACTIVITY

Flexor spasms and extensor spasms are well recognized, particularly in patients with spinal cord injury. These appear to be due to exaggeration of existing flexor and extensor reflexes.

Spastic dystonia is a term coined by Denny-Brown to explain sustained involuntary contractions of muscles that were not driven by reflexes in monkeys with upper motor neuron lesions. For example, there might be tonic contraction of elbow flexors in the absence of stretch or of any stimulation that could provoke a flexor spasm. The contraction continued even after section of the relevant dorsal roots indicating that it was not a peripherally driven reflex. However, that does not necessarily indicate that spastic dystonia can not be modified. For example, when the monkeys were inverted, elbow flexion changed into tonic elbow extension. In humans subjects, spastic dystonia can either increase or decrease with tonic stretch.

As in basal ganglia disorders, spastic dystonia can be purely action-induced. For example, a hemiplegic patient can have normal lower limb tone at rest but upon standing and attempting to walk can develop the well-known hemiplegic posture of extension at the knee and plantarflexion and inversion of the ankle. Furthermore, standing and walking can evoke elbow, wrist, and finger flexion.

CONCLUSIONS

The pathophysiology of spasticity has a number of clinical implications:

- Spasticity is a narrowly defined entity and only one of several types of motor overactivity that can occur after an upper motor neuron lesion, which mostly have separate pathophysiological mechanisms. Terminology precision is essential.

- The clinical pattern of motor overactivity is largely determined by the location and extent of the lesion.
INTRODUCTION

Spasticity is only one component of the motor dysfunction resulting from upper motor neuron lesions. Each component of the upper motor neuron syndrome (UMNS) should be addressed to properly manage these patients (see Table 1). Establishing realistic treatment goals with the patient and the interdisciplinary team can optimize outcomes. Obtaining patient assessment of both active and passive function and identifying areas of problematic muscle overactivity can help target which intervention is most appropriate for each patient. Recent treatments now are available to manage muscle overactivity but need to address other components of the UMNS for optimal functional outcomes.

ASSESSMENT

Patient assessment should include both static and dynamic measures to determine an overall treatment strategy. Assessments from physical and occupational therapists, as well as input from the patient and their caregivers, are essential for establishing patient-specific goals and an optimal treatment plan.

Clinical, electrophysiological, and biomechanical measures have been used to quantify spasticity (see Table 2). The most commonly used clinical measures of spasticity are the Ashworth and modified Ashworth scale (see Table 3). The modified Ashworth scale adds an additional intermediate grade (1+), but has less inter-rater reliability than the Ashworth scale. The Tardieu scale (see Table 4) has advantages over the Ashworth scale in that it not only quantifies the muscles’ reaction to stretch but it controls for the velocity of the stretch and measures the angle at which the catch or clonus occurs. The “spasticity angle” is a neural component that is amenable to treatment.

Electrophysiologic tests such as the H-reflex, H/M ratio, F wave, and tonic vibration reflex (TVR) have used to been used to quantify spasticity. Historically, these measures tend to correlate poorly with the degree of spasticity. The soleus H/M ratio can be used to confirm the effects of intrathecal baclofen (ITB) during the ITB trial and potentially for troubleshooting. In a prospective case series

<table>
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<th>Table 1 Upper motor neuron syndrome</th>
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<tr>
<td><strong>Positive symptoms</strong></td>
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<td>Spasticity</td>
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<tr>
<td>Spastic co-contraction</td>
</tr>
<tr>
<td>Spastic dystonia</td>
</tr>
<tr>
<td>Flexion/extension synergistic muscle patterns</td>
</tr>
<tr>
<td>Reflex release phenomena</td>
</tr>
<tr>
<td><strong>Negative symptoms</strong></td>
</tr>
<tr>
<td>Weakness, fatigue</td>
</tr>
<tr>
<td>Loss of dexterity, balance</td>
</tr>
<tr>
<td>Loss of selective muscle control</td>
</tr>
<tr>
<td><strong>Rheologic changes</strong></td>
</tr>
<tr>
<td>Contracture, fibrosis, atrophy</td>
</tr>
</tbody>
</table>
stroke patients, 17 traumatic brain injured patients, and 4 anoxic brain injured patients treated were with a 50 µg bolus of ITB. Five hours post bolus there was a reduction of the H/M ratio from 62% ± 28% to 14% ± 19% and the Ashworth score decreased from 2.4 ± 0.7 to 1.5 ± 0.6 on the more involved side. This study suggests that the soleus H/M ratio may be more sensitive than the Ashworth score in detecting a physiologic response to an ITB bolus.

Biomechanical measurements of spasticity using a servo-controlled motorically driven device can provide a more reliable measure of spasticity but is limited to the research laboratory. The device can

provide a controlled stretch of a limb while measuring torque, joint angle, and reflex electromyographic (EMG) activity.

The assessment of patients with UMNS should also include measures of motor control (see Table 5). The Fugl-Meyer scale is a reliable and validated measure of upper and lower extremity motor impairment based on the natural progression of functional return after a stroke. The Fugl-Meyer scale has been demonstrated to have high intra-rater and inter-rater reliability, and it can be completed in 10-20 minutes. Decline of function on the Fugl-Meyer scale has been shown to correlate closely with the severity of spasticity. The functional test for the hemiparetic upper extremity was developed at Rancho Los Amigos Hospital and consists of 17 graded tasks with seven levels of difficulty.

Table 2 Measures of spasticity

<table>
<thead>
<tr>
<th>Clinical</th>
<th>Ashworth scale</th>
</tr>
</thead>
<tbody>
<tr>
<td>Modified Ashworth scale</td>
<td></td>
</tr>
<tr>
<td>Tardieu scale</td>
<td></td>
</tr>
<tr>
<td>Oswestry scale</td>
<td></td>
</tr>
<tr>
<td>Spasm frequency scale</td>
<td></td>
</tr>
<tr>
<td>Tone assessment scale</td>
<td></td>
</tr>
<tr>
<td>Electrophysiological</td>
<td>H/M ratio</td>
</tr>
<tr>
<td>Biomechanical</td>
<td>Servo-control device</td>
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</table>

Table 3 Ashworth scale

<table>
<thead>
<tr>
<th>Score</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>No increased tone</td>
</tr>
<tr>
<td>1</td>
<td>Slight increase in muscle tone, manifested by a catch and release or by minimal resistance at the end of the range of motion when the affected part(s) is moved in flexion or extension</td>
</tr>
<tr>
<td>2</td>
<td>More marked increase in muscle tone through most of the range of motion, but affected part(s) easily moved</td>
</tr>
<tr>
<td>3</td>
<td>Considerable increase in muscle tone, passive movement difficult</td>
</tr>
<tr>
<td>4</td>
<td>Affected part(s) rigid in flexion or extension</td>
</tr>
</tbody>
</table>

Table 4 Tardieu scale

<table>
<thead>
<tr>
<th>Quality of muscle reaction</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>No resistance</td>
</tr>
<tr>
<td>1</td>
<td>Slight resistance</td>
</tr>
<tr>
<td>2</td>
<td>Catch followed by a release</td>
</tr>
<tr>
<td>3</td>
<td>Fatigable clonus (&lt; 10 s)</td>
</tr>
<tr>
<td>4</td>
<td>Infatigable clonus (&gt; 10 s)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Angle of muscle reaction (spasticity angle)</th>
<th>Equation</th>
</tr>
</thead>
<tbody>
<tr>
<td>= Angle of arrest at slow speed V1 – Angle of catch as fast speed V3</td>
<td></td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>Velocity of stretch</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>V1</td>
<td>As slow as possible</td>
</tr>
<tr>
<td>V2</td>
<td>Speed of limb falling under gravity</td>
</tr>
<tr>
<td>V3</td>
<td>As fast as possible</td>
</tr>
</tbody>
</table>

Dynamic polyelectromyographic (PEMG) recordings can be used to identify the timing and duration of muscle overactivity in poststroke spastic hypertonia. PEMG recordings can be helpful for understanding muscle involvement for potential treatment with chemodenervation, chemical neurolysis, or surgical release of individual muscles. For example, in the patient with a spastic flexed elbow, PEMG can be used to identify spastic co-contraction of the elbow flexors and extensors. By combining PEMG with kinematic data from a motion analysis laboratory, the primary upper or lower extremity motor dysfunction can be localized. Quantitative gait analysis can differentiate quadriceps overactivity from hip flexor weakness or poor ankle mechanics as the cause of stiff-legged gait.

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Table 5 Measures of motor control

<table>
<thead>
<tr>
<th>Clinical</th>
<th>Upper extremity test (Rancho, Wolf)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Electrophysiological</td>
<td>Dynamic electromyography</td>
</tr>
<tr>
<td>Biomechanical</td>
<td>Motion analysis systems</td>
</tr>
</tbody>
</table>

INTERVENTIONS

Prior to any intervention the goals of treatment should be established. Goals for spasticity management include symptomatic relief or reduced impairments and improvement in active or passive function. Once the patient goals are established, complimentary treatments can be initiated (see Table 6). Certain conditions that can increase spasticity should be addressed prior to initiating additional treatment (see Table 7).

FOCAL/SEGMENTAL TREATMENTS

Chemodenervation with botulinum neurotoxin (BoNT) and chemical neurolysis with phenol can be used alone or in combination to effectively manage focal spasticity. Phenol is reviewed elsewhere in this course. BoNT causes a chemical denervation by inhibiting the release of acetylcholine (Ach) from the peripheral cholinergic nerve
endings. When injected intramuscularly, these toxins cause focal denervation and a graded muscle weakness. Numerous studies and metaanalysis support the use of BoNT in the treatment of focal spasticity. The dose is adjusted based on patient weight, muscle size, and desired effect. The duration of effect depends on dose, dilution, size of the muscle, injection technique, and postinjection therapy. On average, the duration of effect is usually 3-4 months. The most common side effects are adjacent muscle weakness, pain with injection, hematoma, and transient fatigue or nausea. BoNT can be injected by muscle palpation, with needle EMG guidance, low-intensity electrical stimulation or ultrasound guidance. More study is needed to delineate which technique is most effective.

Surgical procedures are typically reserved for those patients with muscle or tendon shortening who have not responded to the less invasive procedures or as a last resort. Tendon transfer, release, or lengthening are the most common orthopedic procedures. The split anterior tibial tendon transfer (SPLATT) and tendon Achilles lengthening (TAL) have been used to manage the spastic equino-varus foot. In a 1-year study of 21 stroke patients 1 year after a SPLATT, 83% reported good or excellent results. All ambulatory patients had improved gait and 35% were able to discontinue their orthosis. Poor surgical outcomes were associated with nonambulatory status. Eighteen patients with spastic hand deformities had improved prehension after release of the flexor–pronator origin and step-cut lengthening of flexor pollicis longus. PEMG may be useful in identifying which patients would benefit most from orthopedic procedures.

### Table 6 Complimentary treatments

<table>
<thead>
<tr>
<th>Rehabilitation treatments</th>
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<tbody>
<tr>
<td>Reduce nociceptive input</td>
</tr>
<tr>
<td>Focal/segmental treatments</td>
</tr>
<tr>
<td>Nerve/motor point blocks</td>
</tr>
<tr>
<td>Tendon transfer/lengthening</td>
</tr>
<tr>
<td>Generalized treatments</td>
</tr>
<tr>
<td>Oral/intrathecal medications</td>
</tr>
<tr>
<td>Rhizotomy</td>
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### Table 7 Factors that increase spasticity

<table>
<thead>
<tr>
<th>Infections: urinary tract infections, pneumonia, cellulitis</th>
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<tbody>
<tr>
<td>Bowel impaction</td>
</tr>
<tr>
<td>Kidney stone</td>
</tr>
<tr>
<td>Deep venous thrombosis</td>
</tr>
<tr>
<td>Ingrown toe nails</td>
</tr>
<tr>
<td>Changes in temperature</td>
</tr>
<tr>
<td>Psychological factors</td>
</tr>
<tr>
<td>Diet, medications</td>
</tr>
<tr>
<td>Disease progression</td>
</tr>
</tbody>
</table>

### Table 8 Pharmacologic treatments

<table>
<thead>
<tr>
<th>GABA system</th>
<th>Baclofen, benzodiazepines</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ion Flux</td>
<td>Dantrium, gabapentin</td>
</tr>
<tr>
<td>Monoamines</td>
<td>Tizanidine, clonidine</td>
</tr>
</tbody>
</table>

| GABA = gamma-aminobutyric acid |

ITB has become a widely used method for the management of intractable spasticity of cerebral or spinal origin. ITB can be safe and effective in hemiparetic spastic patients as well as quadriparetic and paraparetic patients. The advantages of ITB include the procedure's reversible, the noninvasive dose adjustments, its potential for fewer side effects than oral drugs, the evidence to support efficacy in reducing spasticity, and its potential to improve function, comfort, and care. Potential side effects include hypotonia, somnolence, nausea/vomiting, headache, dizziness, and paresthesias. Catheter and procedural complications may occur. Overdose rarely happens. Baclofen withdrawal is more common and can be a life-threatening situation.

### Rehabilitation Treatments

Collaboration with physical and occupational therapists is essential to the effective management spasticity. The therapist can help with patient evaluations, education, and establishing patient specific goals. The primary focus of these therapies is aimed at providing prolonged stretch to shortened muscles and tendons, reducing muscle overactivity, strengthening weak muscles, and improving motor control (see Table 9). Unfortunately, there is a significant variation in the use of these therapies and a lack of controlled trials.

### Table 9 Rehabilitation interventions

<table>
<thead>
<tr>
<th>Inhibitory/serial casting</th>
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<tbody>
<tr>
<td>Weight bearing</td>
</tr>
<tr>
<td>Neurofacilitatory techniques</td>
</tr>
<tr>
<td>Electrical stimulation</td>
</tr>
<tr>
<td>Aquatic therapy</td>
</tr>
<tr>
<td>Electromyographic biofeedback</td>
</tr>
<tr>
<td>Constraint-induced movement therapy</td>
</tr>
<tr>
<td>Robotic training</td>
</tr>
<tr>
<td>Partial weight support treadmill training</td>
</tr>
</tbody>
</table>
Surface electrical stimulation has been shown to reduce muscle atrophy, enhance strength, and possibly reduce spasticity. Unfortunately, most studies have shown limited functional improvement and many patients are unable to tolerate the electrical stimulation. A new device combines a wrist–hand orthosis with functional electrical stimulation (FES) of wrist and finger flexors and extensors during a repeated prehension exercise paradigm. The NESS H200™ is a neuroprosthesis that is well tolerated and has been shown to reduce spasticity and improve function.24 Similar commercially available devices for the leg are the NESS L300™ and the WalkAide. Although these devices are well tolerated and effective more study is needed.

CONCLUSION

Comprehensive spasticity management is essential to optimizing functional outcomes. Careful patient assessment, realistic goals, knowledge of the functional peripheral anatomy, and technical skills are essential to safely and effectively manage spasticity.

REFERENCES

INTRODUCTION

Spasticity is defined clinically best as: Velocity-dependent increase in resistance to passive stretch of a muscle or a group of muscles when a joint is moved passively. This phenomenon is explained by quantitative measurement of spasticity by Tardieu (see Fig. 1). In that figure, to quantify the degree of spasticity of knee flexor, the ankle joint is dorsiflexed passively at a slow pace until the resistance is felt, the angle of dorsiflexion (R2) is measured at that position, then the same movement is repeated at a faster rate and the angle at which the resistance is felt first (R1) is noted. The difference in the angle R1 and R2 is primarily due to dynamic spasticity. This method not only diagnoses spasticity but also helps in distinguishing pure spasticity from other reasons of hypertonia—rigidity, contracture, and dystonia. Early spasticity can be present only during active limb movement, whereas passive movement may not show increased resistance. For example, on passive abduction of the hips in supine posture, an infant may not demonstrate increased muscle tone, whereas lifting up the infant suddenly, holding at the axilla, may produce hip adduction in the form of scissoring. This concept helps in identifying the spasticity at an early stage. Another example is in a child with spastic diplegia. Formal muscle tone examination at lying posture may be normal, but while walking one can observe the spasticity kicking in, thus affecting the walking.

Spasticity is produced as part of the upper motor neuron syndrome, classically defined as the involvement of the neuraxis from the cortical motor neuron to the lower motor neuron (anterior horn cell at the spinal cord or motor cranial nerve nuclei at the brain stem). Figure 2 demonstrates the upper and lower motor neurons. Though the concept of development of spasticity secondary to upper motor neuron lesion is very useful to understand various management options of spasticity, it is too simplistic, and, the underlying mechanism is much more complex. Pure corticospinal or corticonuclear lesion does not produce spasticity, rather, it produces hypotonia. Lesions of other motor tracts—vestibulospinal, reticulospinal, and rubrospinal—in varying combinations may underlie development of increased muscle tone in the form of spasticity. Whatever the mechanism, this diagram simplistically helps in understanding the mechanism of various modalities of spasticity management. So far, spasticity management is concerned with the management of hyperactivity of the lower motor neuron starting from the anterior horn cell to the anterior nerve root to peripheral nerve to muscle and from the muscle sensory organs (spindles, Golgi tendon organs) to the sensory nerve to the dorsal root and finally synapsing with the anterior horn cell, thus completing a monosynaptic reflex arc. This hyperactivity of the lower motor neuron, no more under inhibitory control of the upper motor neuron explains most of the features with spasticity. Thus far, most of the management options are based on controlling this lower motor neuron hyperactivity. Unless therapies such as stem cell therapy prove effective in dealing with primary pathology in brain or spinal cord, physicians have very little control over the upper motor neuron, the primary site of pathology in disorders producing spasticity. Ongoing physical and occupational therapies may help in the recovery or regeneration process through new synapse formation, new nerve endings, and activation of the dormant areas to take over the function of the damaged areas through the process called plasticity. Thus physical and occupational therapies (PT/OT) and orthotic management are the default in the management program for spasticity. Whatever
Spasticity management is proscribed these are absolutely essential, delivered either formally under supervision or through caregivers after teaching them.

Spasticity may not be parallel with weakness. This is an important point to consider before deciding to begin spasticity management. Sometimes spasticity may act as a splint to support the limb in the presence of weakness; as example, quadriceps spasticity may help in ambulation in a child with spastic diplegia. In that case controlling quadriceps spasticity may expose underlying weakness and thus a loss of ambulation. Therefore, clinicians must determine a goal prior to embarking on the spasticity management. On the contrary, in another child with spastic diplegia, ankle planteflexors, hamstrings, and/or hip adductors may be too tight to act as mechanical hindrance to ambulation. In these situations, lowering the excess muscle tone in those selected muscle groups will help the gait and may prevent falls, making the walking more energy efficient.

Spasticity management in children is a challenging problem as the disorders with spasticity have many complications and comorbidities involving multiple systems. For example, a child with a spastic quadriplegic form of cerebral palsy may have seizures and behavioral problems requiring pediatric neurological expertise. There may be contracture of spastic muscles leading to muscle shortening with consequent joint and bony deformities requiring pediatric orthopedic expertise. Appropriate PT/OT and orthotic measures are absolutely essential in management of any child with spasticity. In addition to ongoing effective PT/OT and use of orthotics when needed, the specific management can be broadly categorized (see Table 1) as:

1. Oral medications in the form of baclofen, benzodiazepines, tizanidine, and dantrolene. Each of these medications has unique usefulness and a side effect profile and should be managed by a clinician familiar with them.
CAUSES OF SPASTICITY IN CHILDREN

Table 2 shows various causes commonly responsible for development of spasticity in children arranged in order of the most common. Disorders with spasticity in children alternatively can be classified as: 1) static—secondary to remote brain or spinal cord injury or stroke, hypoxia/ischemia, treated brain tumor, congenital brain malformation, treated hydrocephalus, meningitis or encephalitis with complications, or 2) progressive—secondary to leukodystrophies, hereditary spastic paraplegias, rarely multiple sclerosis, late manifestation of neuronal storage disorders, untreated hydrocephalus, and nontreatable brain tumor. This distinction is very important for a long-term management approach. Also important is the comorbidities that may be relative or absolute contraindication to some surgical management or botulinum toxin injection. The common comorbidities are listed in Table 3.

EVALUATION OF SPASTICITY IN CHILDREN

Modified Ashworth tone scale evaluates the resistance (degree of force) needed to move a joint passively. This is a very easy-to-use bedside evaluation of spasticity with the following limitations: 1) it becomes unreliable in the presence of dystonia; 2) it does not distinguish between various causes of hypertonias such as dystonias, rigidity, contracture, voluntary resistance, muscle spasm (so the clinician has to be extremely cautious about its implication in the clinical context); and 3) it does not reflect the function.
The Tardieu method possibly may be more reflective of the actual degree of spasticity. The principle behind this method is the velocity dependence of the spasticity. However, (as shown in Fig. 1) this method is a bit cumbersome, requiring measurement of the angles of the joint. When a joint is moved passively slowly, the angle of the joint at the maximum range of movement (R2) is measured and this angle is compared to the angle (R1) when the joint is moved faster, thus causing the spasticity to emerge far before the R2 angle. The difference between these angles can be extrapolated to be due to spasticity. This method appears to be very useful in focal or multifocal spasticity for monitoring the treatment response, too. For quantification of spasticity, a simple hand-held dynamometer can measure the force needed to move a joint against resistance.

Hip adductor angle, popliteal angle, and determination of range of movements of various joints, both active and passive, are good ways of assessing the stiffness due to spasticity or other causes mentioned earlier.

From a functional perspective, the functional score of each limb (0-100), the disability score (0-4), the spasm score, the pain score are also simple but useful tools. For children with cognitive problems or for younger children, the parents can be asked the questions to determine the scores. Time to walk 25 ft in ambulatory child is another functional measure.

When the whole child is considered in the context of all the comorbidities (Table 3), the Gross Motor Functional Classification System is a handy way to assess motor functional status, the Functional Independence Measure for Children (Wee-FIM) is a good measure of overall functional abilities, and activities of daily life (ADL) by Barthel is a good alternative measure. The Pediatric Quality of Life Index (PQLI) may reflect the effect of intervention on spasticity. The Goal Attainment Scale may be another way to follow the treatment response. The details of the these methods are beyond the scope of this discussion.

### REASONS TO TREAT SPASTICITY

Each child with spasticity should be assessed individually by a multidisciplinary team, as mentioned earlier. This team should work closely with the parents or care givers to determine the goals of spasticity management in each case. The following are some broader reasons to treat spasticity:

1. **Improved ambulation and greater independence** which may improve the child’s confidence to help them develop their full potential.

2. **Ease of care** in nonambulatory children which may vary from easy transfer, dressing, and undressing.

3. **Improved perineal, axillary, and palmar hygiene** through better stretching and cleaning those areas which are prone to infection.

4. **Prevention and/treatment of painful spasms**, either spontaneous, nocturnal, or during stretching, thus mitigating the suffering of the child and improving their quality of life.

5. **Improvement of range of movement**, prevention or delaying develop in of contractures or bony or joint deformities, thus delaying or preventing the need for major orthopedic interventions such as tendon lengthening, tendon transfer, or osteotomies. In this context it is worthwhile to remember that orthopedic interventions are an absolute necessity in some children, but, on principle, these are performed not to treat spasticity but to treat the complications of unmanaged spasticity either due to the biological reasons or due to lack of proper early institution of spasticity management.

### Consequences of Untreated Spasticity

The consequences of untreated spasticity include:

- Disuse atrophy and weakness of the muscles
- Development of muscle/tendon or soft tissue fibrosis or contracture
- Secondary bony or joint deformities
- Painful muscle spasm/pain
- Poor hygiene and consequent infection
- Problems in caregiving

### VARIOUS OPTIONS FOR SPASTICITY MANAGEMENT IN CHILDREN

**Selective Dorsal Rhizotomy**

In selective dorsal rhizotomy, lumbar laminectomies are performed followed by exposure of the dorsal nerve roots in the spinal canal. They are dissected very cautiously to isolate individual rootlets, then individual rootlets are electrically stimulated to determine whether they are hyperactive. For assessing the hyperactive rootlet, each rootlet is stimulated by a needle electrode with concomitant EMG recording of various lower limb muscles. If the effect spreads to other segments either on the same side or other side, or the effect in the same root-innervated muscles is much prolonged even after a brief single stimulus, the rootlet is considered hyperactive and then severed. Thus, this procedure requires a prolonged careful microsurgery in which an experienced neurosurgeon works together with an experienced electrophysiologist. Needle EMG recordings during the rootlet stimulation are shown in Figures 3 and 4. Figure 3 shows a normal response after right L4 rootlet stimulation, whereas Figure 4 shows an hyperactive left L3 rootlet.

This is a permanent procedure which requires prolonged intensive physical therapy with extensive stretching and strength training for
A year after the surgery. Ideal candidates are children with cerebral palsy, aged 3-10 years with normal cognition, and good lower limb muscle strength. The benefits are 1) segmental/subsegmental—decreased spasticity, improved strength on intensive therapy, improved mobility, and persistent improvement over more than 10 years; and 2) suprasegmental—improved speech pattern, oropharyngeal control, improved fine motor control, and even improved cognition. The mechanism behind this suprasegmental improvement is unclear. This may be related to general improvement in the well being of the child.

Table 4 shows the pros and cons of selective dorsal rhizotomy.

MEDICAL MANAGEMENT OF SPASTICITY IN CHILDREN

Children may tolerate oral or systemic medication better than adults. The medications commonly tried are baclofen, benzodiazepines, dantrolene, and, rarely, tizanidine. The general principle is to start the medications at as low as possible and then very slowly escalate the dose. The night-time dose should be increased first to mimic physiologic sleep awake state.

**Benzodiazepines**

Benzodiazepines act at presynaptic receptors linked to gamma-aminobutyric acid (GABA)-A receptors and alter the chloride channel. As GABA-A receptors are widely distributed, these medications have more central nervous system side effects such as drowsiness, generalized weakness, ataxia, memory impairment, and cognitive issues. There may be issues with drug abuse and withdrawal symptoms with benzodiazepines. Rarely, there may be paradoxical aggression. The most common one used is diazepam—both enteral and injectable preparations are available. So the oral route easily can be changed to intravenous if an oral or enteral intake is not allowed due to any reason. The usual range of oral dose of diazepam is 0.2-0.8 mg/Kg/day. Clonazepam is available primarily in enteral form, the maximum dose is up to 3 mg/day. The side effect profile is similar to that of diazepam.

**Dantrolene**

Dantrolene primarily acts on the muscle membrane and inhibits calcium release from the sarcoplasmic reticulum. Hypertonia, hyperreflexia, and range of movement scissoring may improve but
Tizanidine

Tizanidine is an alpha-2 adrenergic agonist which is beneficial for muscle spasticity and spasm. The side effect profile is also sedation and tiredness. Efficacy is modest at best. It can be used as adjuvant along with other antispasticity medication or modalities of management.

Baclofen

Chemically baclofen is 4-chlorophenyl-gamma aminobutyric acid; as obvious from Figure 5, this is GABA-mimetic, primarily on the GABA-B receptor. It inhibits supraspinal excitatory and hyperactive spinal cord synapses through inhibition of calcium influx into the presynaptic terminal, thus reducing presynaptic excitatory neurotransmitter release. Baclofen also inhibits substance P and some dopamine neurons. This may explain the analgesic effect of baclofen in addition to an antispasticity effect. Baclofen does not have any intravenous formulation. After oral administration it is rapidly absorbed, partially metabolized in liver, and largely excreted unchanged in the kidneys. The dose is not that closely related to age or weight. Oral dose of 30-90 mg will result in cerebrospinal (CSF) concentration of 12-95 ng/ml, whereas an intrathecal dose of 400 mcg is associated with a CSF concentration of 380 ng/ml. That is why the concept of intrathecal baclofen pump came up. Side effects of oral baclofen are dose related—excessive hypotonia and weakness, impaired cognition, confusion, memory and attention difficulties, and, rarely, paradoxical excitation and insomnia.

Intrathecal Baclofen Pump

To deliver the medicine directly around the site of its action, baclofen can be injected at the spinal subarachnoid CSF space through a programmable, implantable pump delivering the medicine continuously. The rate of infusion can be varied and programmed through a handheld computer (programmer). The intrathecal baclofen (ITB) pump is refilled at intervals by a transcutaneous injection. Figure 6 diagramatically shows the pump system. The pump lying inside the parietal abdominal wall is connected via a catheter to the CSF space entering through the lumbar area, the tip is then advanced upwards to the desired segmental level where the spasticity is at its maximum. The indication is that at least a moderate degree of spasticity responds well to oral baclofen but systemic side effects of sedation and cognitive issues develop. Severe scoliosis, poor trunk muscle strength, and the presence of a

### Table 4 Pros and cons of selective dorsal rhizotomy

<table>
<thead>
<tr>
<th>Pros</th>
<th>Cons</th>
</tr>
</thead>
<tbody>
<tr>
<td>One time procedure; may be an advantage to families with poor availability of medical care.</td>
<td>Prolonged surgery requiring expert neurosurgeon and electrophysiology team.</td>
</tr>
<tr>
<td>No implanted hardware, thus risk of infection is minimized; no recurring complication.</td>
<td>Permanent procedure with no adjustment possible later; if too many rootlets are cut, there may be permanent weakness and sensory or bladder involvement, and, if too few are cut, it will not be effective.</td>
</tr>
<tr>
<td>If ineffective or effectivity is lost later, can proceed to other modalities such as baclofen pump or botulinum toxin injection.</td>
<td>Needs committed extensive physical therapy to achieve gain; if not the weakness may be more overt.</td>
</tr>
</tbody>
</table>

with limited objective functional improvement. The limiting side effects are drowsiness, weakness, nausea, diarrhea, and, most importantly, hepatotoxicity (1.8%).
concomitant ventriculo-peritoneal shunt and other hardware inside the body are relative contraindications for the ITB pump. For the best case selection it is recommended to have a trial of ITB in which the children are subjected to objective evaluation before and serially after ITB injection. The pros and cons of the ITB pump have been highlighted in Table 5. The major complications are highlighted in the Table 6. Regarding scoliosis progression following the ITB pump, current literature does not support such relationship. The cost-effectiveness of ITB pump has been well analyzed by Edgar and colleagues (2007). The incremental cost per quality-adjusted life-year for identical cohorts of children treated with an ITB pump was analyzed. For the ITB group the 5-year cost of treatment increased by $49,000 relative to alternative treatment. However this was accompanied by average gain of 1-2 quality-adjusted life-years. The net result was an incremental cost-effective ratio of $42,000 per quality-adjusted life-year, below the $50,000 to $100,000 range, accepted as very cost-effective.

Selective Dorsal Rhizotomy or Intrathecal Baclofen Pump?

Significant involvement of the upper limbs and cognitive/behavioral affection will contraindicate rhizotomy. For candidacy of the ITB pump the body weight should be preferably above 25 lb, or there may be a mechanical problem implanting the pump in the parietal abdominal wall. The presence of overt weakness in the muscles, even if there is primary lower limb involvement with preserved cognition, is a contraindication for the dorsal rhizotomy. For children below 3 years or above 10 years of age rhizotomy is not recommended. Associated movement disorder may be against the rhizotomy; whereas dystonia, if present, may respond well to an ITB pump.

**Figure 6** Intrathecal baclofen pump system.

Source: http://www.clevelandclinic.org/health/health-info/pictures/pumpn-bdy.gif

**Table 5** Pros and cons of intrathecal baclofen pump

<table>
<thead>
<tr>
<th>Pros</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adjustable and reversible effects</td>
</tr>
<tr>
<td>Less invasive than surgery</td>
</tr>
<tr>
<td>Longterm reduction in spasticity</td>
</tr>
</tbody>
</table>

**Cons**

- Implanted device
- Frequent return visits—refills, adjustments, and new batteries
- Risk of infection and catheter malfunction
- Baclofen withdrawal can be life threatening
- Requires compliance

**Table 6** Intrathecal baclofen pump complications in adults and children

<table>
<thead>
<tr>
<th>Procedures over 5 years</th>
<th>Revision of pump (%)</th>
<th>Catheter revision (%)</th>
<th>Infection (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adult</td>
<td>72</td>
<td>2.7</td>
<td>14</td>
</tr>
<tr>
<td>Pediatric</td>
<td>146</td>
<td>3.4</td>
<td>24</td>
</tr>
</tbody>
</table>

**Botulinum Toxin Injection**

Botulinum toxin is produced by the bacteria Clostridium botulinum. Among seven neurotoxins produced by various serotypes, botulinum toxin type A (Botox®, Dysport®) and type B (Myobloc®) are purified to be used as one of the safest management options for spasticity and/or dystonia among many other conditions including drooling of saliva which is a very common comorbid condition associated in spastic children with cerebral palsy. The chemical structure of the botulinum toxin has been shown in Figure 7; it has two chains—a light and a heavy chain connected by disulfide bridge. Figure 8 (on page 17) shows the mechanism of action of botulinum toxin. The molecule enters the presynaptic junction at the neuromuscular junction, then is cleaved at the lysosome. The light chain is the active compound which cleaves the docking proteins, synaptobrevin or SNAP-25, which normally helps fusion of the acetylcholine vesicles to the presynaptic membrane to release acetylcholine in the neuromuscular junction. This produces an action potential that propagates along the muscle membrane. This toxin is very effective for focal or multifocal spasticity and/or dystonia and also is used effectively in the focal component of a generalized spasticity or dystonia. When compared to neurectomy and dorsal rhizotomy, the effect of a botulinum toxin injection is reversible. This reversibility is however a double-edged sword as the effect also is reversed along with any side effects. So, there is need for reinjection every 3-5 months. There is no cognitive or central nervous system side effect. The overall side effect profile is excellent, the response is very predictable. Concomitant salivary gland injection helps drooling significantly.

Efficacy of botulinum toxin has been evaluated by the therapeutic and technology assessment subcommittee of the American Academy of Neurology in 2008 (Table 7). They found six class I studies showing beneficial effect of botulinum toxin for spasticity,
thus their recommendation regarding botulinum toxin in management of spasticity in children is class A (should be offered). However this option cannot be a primary option for management of a generalized spasticity as the dose requirement may be too high. Its use should be reserved in focal or multifocal spasticity, or it can be used to manage the most severely spastic muscles in combination with other modalities of management such as the ITB pump or dorsal rhizotomy. Very recently, the United States Food and Drug Administration has approved the use of botulinum toxin in adults with spasticity. Use in children with spasticity still comes with a black box warning that the family should be alerted to seek immediate medical attention if the child develops dysphagia, dyspnea, dysphonia, or generalized weakness following botulinum toxin injection as part of systemic botulism.

Neurectomy/Neurolysis

Surgical sectioning of a nerve (neurectomy) supplying a single muscle or a group of muscles and phenol or alcohol block of the nerve (neurolysis) are performed very rarely in children with spasticity. The reasons may be irreversibility of the neurectomy and pain involved in neurolysis, possibly further contributed by excellent response to botulinum toxin injections. However in specific situations in which spasticity is severe in a single muscle group, as in the hip adductor, if ambulation or use is not a concern, a decision may be made to perform these procedures even in children.

SPASTICITY MANAGEMENT: CHILDREN VERSUS ADULTS

General Management

The younger developing brain has more plasticity, thus early intervention with therapy is much more beneficial in children. If therapy is delayed or inappropriate there may be misdirected plasticity. Children in general tolerate oral medications better than adults.

Growth spurts in adolescent children with spasticity may increase the muscle and limb length discrepancy, producing more stiffness, more progression of orthopedic deformities, worsening of gait in ambulation, hip dislocation, and bony and joint pain. Thus, more active intervention is needed in adolescence. Scoliosis also progresses rapidly during adolescence, so its management is of paramount importance in addition to other antispasticity management. For example, employing an ITB pump is very challenging when a scoliosis rod is inserted. Additionally, management of spasticity may be challenged by adolescent behavior that results in oft repeated poor compliance in therapies.

There is another important aspect to be kept in the mind when evaluating a child with motor abnormalities following early brain injury. Though the brain pathology may be static, the motor abnormalities may increase. For example, hypotonic cerebral palsy may progress with age to spastic quadriplegic type. Dystonia and some other movement disorders such as chorea and tremors may appear years after after the inciting brain insult. Pseudoregression with increasing motor difficulties during acute stress such as fever and infection elsewhere may lead to a lot of costly investigations if the clinician is not aware of this phenomenon.

Specific Management

The specific management of spasticity in children versus adults includes:

1. Generalized spasticity in children under the age of 3 years, or weight less than 25 lb—PT/OT, modest dose of oral antispasticity medications, added with botulinum toxin in selected muscles, may help buy time for eligibility for dorsal rhizotomy or an ITB pump.
**Figure 8** Mechanism of action of botulinum toxin.

**Table 7** Report of therapeutic and technology assessment subcommittee of the American Academy of Neurology: summary for botulinum toxin in the treatment of spasticity

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Class</th>
<th>No. of subjects</th>
<th>Outcome measures</th>
<th>Adverse events</th>
<th>Conclusions</th>
<th>Recommendations*</th>
<th>Limitations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adult spasticity</td>
<td>14</td>
<td>1 906</td>
<td>Tone (Ashworth), passive fx: range of motion, cleaning, hygiene, pain Active fx: Goal Attainment Scale, Frenchay: global disability (MD/pt)</td>
<td>Focal weakness, pain</td>
<td>Established safe and effective</td>
<td>A</td>
<td>Methodologic challenges in study design</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Probably effective</td>
<td>B</td>
<td>Limited outcome measures to demonstrate efficacy in active functional gains</td>
</tr>
<tr>
<td>Childhood spasticity in cerebral palsy</td>
<td>6</td>
<td>376</td>
<td>Tone (Ashworth), passive fx: range of motion, active fx: gait/video/kinematic analysis; global disability (MD/pt)</td>
<td>Pain, weakness, falls, incontinence, dysphagia</td>
<td>Established safe and effective</td>
<td>A</td>
<td>Best evidence for equinus virus</td>
</tr>
</tbody>
</table>

*Classification of recommendations is available on the Neurology® website at www.neurology.org.

A = should be offered, B = should be considered, C = may be considered, fx = function, MD = physician, pt = patient

From: Simpson DM and colleagues.¹
2. Worsening of spasticity after years of dorsal rhizotomy: must rule out new onset superimposed dystonia; an ITB pump may be considered at that point.

3. Spasticity following hemispherectomy (primarily upper limb, hemineglect, weakness combined), PT/OT, constraint-induced movement therapy, and botulinum toxin injection are the best combination.

4. Children with spasticity and ventriculoperitoneal (VP) shunt—one should be very cautious in employing an ITB pump as there is an increased risk of infection and also possible baclofen overdose.

CONCLUSION

Children with spasticity can be managed very effectively through a multimodality approach managed by multidisciplinary team composed of a child neurologist, neurosurgeon, orthopedican, therapists, and psychiatrists. PT/OT is a must in each case. When therapy alone is unable to control spasticity, one should combine oral medication with a botulinum toxin injection in selected muscles to gain maximum control of spasticity with tolerable side effects. An ITB pump can be effectively combined with botulinum toxin. An ITB pump can be implanted after dorsal rhizotomy if spasticity gets worse with time. Combining orthopedic and neurosurgery procedures may be convenient for the patient and family.

SUGGESTED READING

INTRODUCTION

Doppler was one of the first physicians to use phenol as a neurolytic agent. In 1925, he painted phenol on rabbit ovarian vessels and noted subsequent vasodilatation. Eight years later, in 1933, the Russian literature reports Nechaev used phenol as a local anesthetic agent. Mandle, in 1950, reported the successful injection of phenol for permanent sympathectomy in 15 patients without complications. Both Haxton and Boyd simultaneously reported paravertebral injections of phenol for peripheral vascular disease in 1949. In 1955, Maher reported the use of phenol by intrathecal injection for intractable cancer pain.²⁴ Nathan²⁸ along with Kelly and Gautier-Smith,¹⁹ all simultaneously reported intrathecal injections of phenol for relief of spasticity in 1959.

INTRATHecal PHENOL FOR SPasticITY: HISTORICAL USE

As mentioned above, the earliest uses of phenol for relief of spasticity was by the intrathecal method.¹³,¹⁹,²³,²⁸,³¹ These early studies utilized up to 20% phenol solutions in glycerol. A major concern with the use of phenol intrathecally included significant complications such as loss of residual motor function or sensation as well as loss of sphincter control. Weaker solutions of 5% phenol in glycerin produced effects of shorter duration; however, the 5% phenol solution intrathecally still produced considerable side effects related to the sphincters. It did not appear that phenol caused any noteworthy effect on the cerebrospinal fluid, and no cases of meningitis were reported.

There was significant interest in the specific effects of phenol on neural tissue used intrathecally. Iggo and Walsh¹⁷ studied the effects on spinal roots of cats and concluded that small fibers were more susceptible to phenol than larger fibers. Nathan and Sears²⁹ also looked at the effect of phenol on spinal nerve roots of cats. The study found that: 1) phenol had a temporary local anesthetic effect in addition to its longer term effects on spasticity, 2) aqueous phenol solutions could cause reversible or irreversible block of nervous conduction in both myelinated and unmyelinated nerve fibers, 3) the smallest fibers were blocked before larger fibers, and 4) recovery is most likely to occur in the fastest conducting fibers of the “A” group and are least likely to occur in the smaller “C” fibers. Histopathological changes after the injection of 7.5% to 15% phenol solutions in glycerol in the lumbosacral subarachnoid space of cats show that there was damage to myelin sheaths and axons, as well as vasculitis, arachnoiditis, and peripheral demyelination, in addition to occasional infarction of the lower spinal cord.²
PHENOL PERIPHERAL NERVE AND MOTOR POINT BLOCKS

The search for a simpler, safer, and practical treatment of spasticity led Khalili and colleagues to use 2% and 3% aqueous phenol solutions for selective peripheral nerve blocks. These injections were used in a diverse group of 20 patients with severe spasticity. The spasticity was secondary to various causes including traumatic spinal cord injury, vascular accidents, cerebral palsy, and transverse myelitis. The minimal amount of phenol used to produce an effective block was 0.5 cc of a 2% phenol solution. The maximum amount was 5 cc's of a 3% solution. Overall the average amount of phenol solution used for each nerve block was 2 cc's of a 2% to 3% solution of aqueous phenol. Phenol motor point blocks were first described by Halpern and Meelhuysen in 1965. They initially performed phenol motor point blocks with 3% aqueous phenol, however, they found that the duration effect was limited and increased the aqueous phenol solution to either 5% or 7%. Because the postinjection observation period was limited, any difference in the longer duration of effects between the 5% and 7% groups could not be determined. Thirty-nine patients were injected for a total of 122 muscles. Improvement of tone was achieved in 114 of 122 muscles.

PATHOPHYSIOLOGY OF PHENOL ON PERIPHERAL NERVES

Looking at the histopathological effects after phenol injections can be important in understanding the specific mechanisms of phenol action. Fusfeld performed needle EMG examinations on a small group of hemiplegic patients approximately 1 month after phenol nerve or motor point blocks were conducted. Of the seven patients, three had needle EMG examinations prior to the administration of the phenol blocks and were found to be free of denervation. One month after the phenol block, five of the seven patients had evidence of denervation. The authors concluded that the alpha motor fibers are not all spared by nerve or motor point phenol blocks.

Khalili and Betts found, that in certain patients, voluntary movement emerged following phenol nerve blocks with no clinical evidence of sensory involvement. This was interpreted as a result of the selective effect of dilute phenol on gamma fibers. In other patients, loss of voluntary motor activity and evidence of sensory impairment had been noticed which could be evidence of a more widespread effect on the peripheral nerve. Glass and colleagues studied needle EMG responses on 20 patients after performing phenol nerve and motor point blocks with either a 3% or 6% phenol aqueous solution. All the patients had needle EMG examinations prior to the phenol injections and there was no evidence of denervation. After injections, virtually all patients who had 6% phenol nerve blocks showed evidence of denervation. On the other hand, almost no one with motor point block at a 3% or 6% concentration or phenol nerve blocks with 3% concentration showed evidence of denervation. The authors suggest that the 3% phenol concentration produces a preferential paralysis of the gamma efficients, without significantly effecting alpha motor activity that is affected by the 6% solution.

HISTOPATHOLOGY OF NERVE AND MUSCLE AFTER PHENOL INJECTION

Several studies looked at the histopathology of the peripheral nerve after phenol nerve block. Different concentrations of phenol, ranging from 1% to 6%, showed no evidence of selective destruction of smaller versus larger fibers. There was evidence of perineural inflammatory cell infiltration, axonal degeneration, and myelin sheath degeneration. The amount of degeneration did appear to be concentration dependent. Wallerian degeneration was seen with neural regeneration.

Halpern reviewed histological studies of muscles in rats and dogs after phenol motor point blocks. The localized points of injection proved to be intramuscular nerves that were near the terminal innervation band, or frequently proximal in the mixed nerves, which includes the motor branch and the muscle afferents, running in the neurovascular bundle in the intrafascicular regions of the muscles. It was found that these phenol motor point blocks caused nerve destruction with secondary denervation and atrophy with muscle necrosis. Both the nerve and muscles showed evidence of regeneration, which was well established by 2 months and almost complete by 3 months.

SPASTICITY MANAGEMENT DECISIONS

Treatment of spasticity must be made after full assessment of the patient, including discussion with any caregivers involved. Multiple issues should be considered, including if the spasticity is severe enough for treatment, and, if untreated, will the spasticity lead to permanent contractures. The patient should be evaluated to determine if the spasticity is generalized or localized. Activities of daily living, such as transfers, seating, driving, or self-care, should be evaluated to see if spasticity is interfering. The patient should be evaluated to assess if there is severe chronic pain that may be alleviated by a spasticity procedure. Finally, is the severity of the spasticity problem worth the potential side effects of a phenol injection procedure?

INDICATIONS AND BENEFITS OF CHEMICAL NEUROLYSIS WITH PHENOL

Chemical neurolysis with phenol should be considered as a treatment for spasticity when other more conservative measures have not succeeded. Ideally, the spasticity to be treated should be localized rather than generalized. There are times when more conservative measures such as oral medications may be contraindicated. For example, significant liver function abnormalities can prohibit use of certain antispasticity medications. Furthermore, not all medications are approved for use in pediatric patients. Medications can also cause significant sedation or deleterious effects on cognition.

Phenol chemical neurolysis may be useful in any stage of recovery. In the acute period, it may help prevent loss of range of motion, such as trying to prevent plantar flexion contractures while in an
intensive care unit. More commonly, it is used in the later stages in the treatment of spasticity.

Chemical neurolysis of individual muscle groups can have more widespread effects by decreasing the stimulus for flexor or extensor synergy patterns. In addition, a decrease in spasticity in an agonist muscle can unmask active voluntary activity in a weaker antagonist muscle group. Thus, in certain cases, after phenol blocks are performed, voluntary motor contraction may be observed that did not appear to exist prior to the nerve blocks.

PHARMACOLOGY OF PHENOL

Phenol or carbolic acid, which was first isolated in 1834, is a benzene ring with one hydroxyl group. Lister introduced it to medicine as an antiseptic in 1867. In its pure state, it is a colorless crystal that melts if heated to 38°C. At concentrations up to 6.7%, it is soluble at room temperature.

Phenol has a local denaturing effect on tissue protein. Furthermore, it has an anesthetic effect, which can be of benefit during the procedure. It is bacteriostatic at a 0.2% concentration and bacteriocidal at a 1% concentration. Eighty percent of phenol is excreted by the kidney. Because phenol is a sclerosing agent, care should be taken not to inject it into vascular structures. Phenol does have systemic toxic effects. A toxic dose is estimated between 8 and 15 g. At toxic levels, tremors and convulsions may be seen. Phenol was initially available in glycerin for intrathecal uses; however, presently it is not to inject it into vascular structures. There is one documented case of a patient developing an arterial block in the upper extremity after receiving a median phenol nerve block requiring an upper-limb amputation.

The documented duration of the effect after phenol motor point blocks using 5% aqueous phenol. In 84% of the patients, the effect lasted 4 months or longer, and in 60% of the patients, the effect lasted 6 months or longer. Repeat injections produced similar responses to the initial injection.

DURATION OF EFFECT

The main adverse reactions that have been seen after phenol motor point blocks have been few and consist almost entirely of mild local pain that lasts for a few days after the injections. Occasionally, some local induration has been noted. This was Halpern’s and Meelhuysen’s experience during a total of 394 muscles injected in 95 patients.

The major complication of phenol nerve blocks discussed in the literature is that of paresthesias. After phenol nerve block, approximately 4% of patients will suffer paresthesias. In the vast majority of cases, the pain or paresthesias lasted from several days to a month. The complications generally did not interfere with any functional gains obtained in the patients. A tight glove or sock was found to relieve discomfort. Paresthesias or dysesthesias are less likely to be encountered in children, with one study showing a 0.4% occurrence. When paresthesias and dysesthesias persist, repeating the phenol nerve block can be curative. A more worrisome complication of paravertebral phenol block is possible diffusion into the subarachnoid space. This can lead to bowel, bladder, or sexual dysfunction when the paravertebral blocks are performed in the lumbar area. Cervical paravertebral blocks are never utilized due to the possibility of respiratory depression if there is any diffusion into the subarachnoid space.

INFORMED CONSENT

Due to the above potential complications, informed consent must be obtained when phenol blocks are performed.

EQUIPMENT

The equipment most commonly used is a portable direct current stimulator with a digital readout of the current intensity. The cathode of the stimulator is connected to a Teflon®-coated needle. With the Teflon coating, only the bevel of the needle is exposed, thus the effect of the stimulation is seen only at the point where the phenol is actually being injected. These needles vary in length, Most commonly a 22 gauge is used; however, other gauges are available. The phenol is drawn up in a syringe and extension tubing is placed between the syringe of phenol and the stimulating Teflon-coated needle. The extension tubing helps prevent movement of the needle when the plunger of the syringe is depressed and the phenol is injected.

PEDiATRIC CONSIDERATIONS

Griffith and colleagues discuss the use of anesthesia in phenol intramuscular neurolysis in young children with spasticity. Chloral hydrate at 30 mg/kg of body weight can be given but must be administered orally 90 minutes prior to the procedure. Griffith and colleagues also discuss general anesthesia as an option for treating young children.
A local anesthetic block prior to a phenol block can give an estimation of loss of function due to loss of spasticity. This also can be helpful if there is any question of the degree of contracture versus spasticity in a potential area to be blocked. A trial local anesthetic block is commonly used when a spastic gait pattern is under consideration for phenol procedures. Sometimes lower extremity spasticity may help gait, and taking away the spasticity may cause deterioration of gait even though the upper extremity requires spasticity management. Since it is possible that the spasticity may help gait, the local anesthetic block could predict potential improvement or deterioration of gait. However, a local anesthetic block as a trial for an ambulatory patient is not always analogous to a phenol block. With a local anesthetic block there is also a sensory block which may affect gait as well.

**SPECIFIC ANATOMIC LOCALIZATION**

**Musculocutaneous Nerve**

In the axillary approach, to inject the musculocutaneous nerve,20 the patient is placed in the supine position and the shoulder is abducted. The shoulder also should be externally rotated with the elbow extended. The axillary artery should be palpated and pushed aside posteriorly. The needle is inserted just behind and beneath the tendon of the pectoralis major. The needle is advanced slowly in the direction of the coracoid process.

**Sciatic Nerve**

The sciatic nerve can be found at a point bisecting a line joining the ischial tuberosity and the greater trochanter.7 With needle manipulation it is possible to get primarily the tibial division or the peroneal division of the sciatic nerve at this point.

**Thoracodorsal Nerve**

To inject the thoracodorsal nerve, the needle is inserted at the midpoint of a line from the apex of the axilla to the inferior point of the scapula. The needle should be angled upwards. Optimally for this procedure, the arm should be abducted to 90 degrees, if possible. The thoracodorsal nerve is fairly superficial and can be found at an approximate depth of 1 to 1.5 cm.

**Tibial Nerve**

To inject the tibial nerve, the patient is placed in the prone position with the leg to be injected resting on a pillow to maintain the knee in approximately 30 to 45 degree of flexion. The needle electrode is inserted at the apex of the popliteal fossa or at the popliteal crease at the level of the knee joint.7

**Obturator Nerve**

The patient is placed in the supine position with the thigh in maximum adduction to inject the obturator nerve. The obturator nerve divides into an anterior and posterior branch. The anterior branch is generally injected first. The adductor longus tendon is palpated at the pubic tubercle. The needle is inserted approximately 2.5 cm distal from the origin at its lateral margin. The needle is directed posteriorly and toward the head. For localization of the posterior branch, the needle is inserted approximately 1 cm deeper.1,7,33

**Paravertebral Lumbar Block**

The paravertebral lumbar blocks can be considered for hip flexor spasticity. The L2, L3, and possibly the L4 nerves need to be injected. The patient is placed in the lateral recumbent position on the side opposite that to be injected with the back flexed. From the level of the iliac crest, which corresponds to L4, all the interspaces are marked off up to L2. The needle is introduced approximately 2.5 cm out from the spinous process. The needle is advanced until bone is reached, which is the transverse process. At this point, the needle is retracted and reintroduced with its point slightly more medially and inferiorly making a 20 to 45 degree angle with the sagittal plane. The needle tip is then advanced slowly until the desired responses of the hip flexors are found.125

**Median Nerve**

The median nerve lies slightly medial to the brachial artery at the elbow. The nerve also lies under the bicipital aponeurosis. A point
2.5 cm distal and anterior to the medial epicondyle of the elbow should be found. The needle is inserted at this point. The brachial artery runs laterally to the median nerve. During the advancement of the needle in this approach, a motor branch to the pronator teres may be encountered and this can be blocked.

**Recurrent Motor Branch of the Median Nerve**

To inject the recurrent motor branch of the median nerve, the technique described for the thumb-in-palm deformity is utilized.\(^{18}\) Two intersecting lines are drawn on the palm. A line is drawn starting from the web space of the thumb parallel to the proximal palmar crease. A second line is drawn parallel to the radial border of the long finger. At the intersection of these two lines the recurrent motor branch of the median nerve enters the thenar mass. At this point, the stimulating needle is inserted.

**Subscapularis Motor Point**

The subscapularis motor point block has been described for the treatment of the painful hemiplegic shoulder.\(^{6}\) To inject the subscapularis, the patient is positioned lying on his/her side with the hemiplegic side up. The affected limb is positioned to produce as much winging of the scapula as possible. At the level of the spine of the scapula, the needle is inserted just under the medial edge of the scapula. The needle is advanced close to the anterior surface of the scapula with the objective of localizing the point of maximal internal rotation of the shoulder with the electrical stimulator.

**Tibialis Anterior and Tibialis Posterior Motor Points**

Both the tibialis anterior and the tibialis posterior muscles may be injected for severe foot inversion. Approximately one third of the way down the tibialis anterior, an anterior approach is taken. Once the tibialis anterior motor points are located, the needle is advanced straight through the interosseous membrane to locate the tibialis posterior motor points.

**COMMON PATTERNS OF SPASTICITY**

The common patterns of spasticity include:

**Lower extremity**
- Great toe extension
  - Can use phenol motor point blocks to the extensor hallucis longus
- Ankle/foot equinovarus
  - Depending on the incriminating spastic muscles, the tibialis anterior and tibialis posterior motor points, and the tibial nerve, injection at the level of the knee may be of benefit.
- Hip and knee flexion
  - For hip flexion spasticity, paravertebral lumbar blocks at the L2, L3, and possibly L4 level may be helpful.
- Thigh adduction
- Anterior and posterior branches of the obturator nerve can be considered. Retrospective analysis of 62 patients has shown benefits with hygiene, pain, spasticity, and range of motion with obturator phenol blocks.\(^{42}\)

**Upper extremity**
- Shoulder internally rotated and adducted, elbow flexed and pronated, wrist and fingers flexed
  - For this upper extremity pattern seen in stroke and brain injury patients, one approach is to start with the thoracodorsal nerve. After this injection, the entire upper extremity pattern may improve and the upper extremity should be reevaluated. If there is still significant shoulder adduction spasticity then the next procedure that can be considered is motor point blocks of the pectoralis major. Then going down the arm, the musculocutaneous nerve can be injected for elbow flexor spasticity, and the median nerve for median nerve distribution spasticity.

**REPEAT PHENOL INJECTIONS**

Phenol blocks may be repeated when the effect diminishes; however, it may be more difficult to perform this localization on repeated attempts.

**ADVANTAGES OF PHENOL COMPARED TO BOTULINUM TOXIN**

The advantages of phenol injections compared to botulinum toxin injections include:
- Much lower cost
- Longer lasting effect (However there are comparative studies that say the opposite.)\(^{39} 40 41\)
- Quick onset with effects observed at time of procedure
- Ease of confirmation of the amount of spasticity reduction at time of procedure
- Ability to block larger territory with a nerve block
- Ability to treat more spastic muscle/nerve territories at one sitting

**DISADVANTAGES OF PHENOL COMPARED TO BOTULINUM TOXIN**

The disadvantages of phenol injections compared to botulinum toxin injections include:
- More potential complications, particularly dysesthesia risk
- More difficult technical procedure requiring greater operator skill and longer procedure time
- Need for compounding or hospital pharmacist with hood to prepare
COMBINING BOTULINUM TOXIN AND PHENOL

There will be occasions when there is consideration to use botulinum toxin to treat an adult or pediatric patient but the amount of botulinum toxin required exceeds the maximal recommended dose. It is easy to use phenol for large proximal muscles, or major nerve territories, and use botulinum toxin for smaller distal muscles. This can be performed at the same time.18

PHYSICAL AND OCCUPATIONAL THERAPY COMBINED WITH PHENOL

The addition of physical and occupational therapy in the overall treatment of spasticity appears to improve outcomes.43 However, this has not been extensively or scientifically evaluated when combined specifically with phenol injections. Similarly, the same can be said for serial casting when spasticity and contracture are present. Again, there is the same paucity of scientific studies combining serial casting with phenol.

CONCLUSION

Spasticity may significantly interfere with seating and positioning. Severe spasticity may also interfere with hygiene, transfers, and other activities of daily living. Chemical neurolysis with phenol has evolved into a useful technique in the treatment of spasticity.

REFERENCES


