Advanced Management of Spasticity

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[Image of a space shuttle launch]
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- **Ileana Howard, MD**
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- **Keith Claffey, MA, APN**
  - has no financial interest to disclose

- **George Gombas, MD**
  - has no financial interest to disclose
Learning Objectives

1. Describe a clinical algorithm for spasticity management
2. Discuss indications for advanced spasticity management- including chemodenervation and intrathecal baclofen pump
3. Recognize cases in which advanced spasticity management improves the patient’s outcome beyond what can be achieved through conservative measures
Spasticity

THE PROBLEM
What is spasticity?

Velocity dependent increase in tonic stretch reflexes with exaggerated tendon jerks

Lance 1980
Spasticity

- **Hippocrates**
  - 460-370 BC
  - spasmos

- **Good**
  - 1829
  - “Spastic wryneck”

- **Little**
  - 1843
  - First detailed description of a spastic condition—Little’s disease
Spasticity

- Charcot
  - 1868
  - “Spastic paraplexie”-MS

- Spasticity and rigidity used interchangeably for years

- Lance’s definition
Spasticity

Today- spasticity is a component of “Upper Motor Neuron Syndrome”

- Weakness
- Loss of dexterity and motor control
- Loss of endurance
- Change in muscle tone
- Hyperreflexia and clonus
- Spasticity
- Babinski sign
Spasticity
Spasticity

Pathophysiology

- Innate flexion and extension patterns are suppressed by the brain by sending inhibitory signals to the alpha motor neurons
- Loss of inhibition to the alpha motor neurons secondary to disease or trauma leads to uninhibited contraction of the muscles
- Uninhibited contraction leads to a cascade of events that may lead to significant morbidity
Spasticity

- Uninhibited contraction of muscles
- Difficulty with movement
- Contractures
- Pain
- Debility
- Immobility
- Ulceration
# Spasticity Measurement

### Objective
- Ashworth, 1964
- Modified Ashworth, 1987
  - Bohannon and Smith
- Tardieu Scale
- Gait speed

### Subjective
- Spasm Frequency Scale
- SCI Spasticity Evaluation Tool (SCI-SET)
- Goal Attainment Scale
- Hygiene scale (hip adductor)
# Ashworth Scale

<table>
<thead>
<tr>
<th>Score</th>
<th>Significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>No increase in muscle 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>1+</td>
<td>Slight increase in muscle tone, manifested by a catch, followed by minimal resistance throughout the remainder (less than half) of the ROM</td>
</tr>
<tr>
<td>2</td>
<td>More marked increase in muscle tone through most of the ROM, 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>
<tr>
<td>Score</td>
<td>Significance</td>
</tr>
<tr>
<td>-------</td>
<td>--------------------------------------------------</td>
</tr>
<tr>
<td>0</td>
<td>No spasms</td>
</tr>
<tr>
<td>1</td>
<td>Spasms when stimulated</td>
</tr>
<tr>
<td>2</td>
<td>Occasional spontaneous spasms</td>
</tr>
<tr>
<td>3</td>
<td>&gt;1 but &lt;10 spontaneous spasms/hr</td>
</tr>
<tr>
<td>4</td>
<td>&gt;10 spontaneous spasms/hr</td>
</tr>
</tbody>
</table>
Spasticity

SPECTRUM OF DISORDERS
Spasticity

Conditions that may exhibit spasticity include

- Spinal cord injury
- Multiple Sclerosis
- Stroke
- Brain injury
- ALS
- Cerebral Palsy
Spasticity in Spinal Cord Injury (SCI)

- Chronic SCI (> 1 yr post-injury): 65-78% (Adams & Hicks, 2005)
  - Likelihood may vary by level of injury
- Potential negative effect on QOL
  - restrict ADLs,
  - inhibit walking & self care
  - result in pain, fatigue
  - contractures, pressure ulcers
- Potential positive effect on QOL
  - assist in sitting/standing stability
  - maintain muscle bulk & strength,
  - preserve venous return
Spasticity in MS

- Prevalence: up to 84% (Rizzo, 2004) patients with MS report some spasticity
  - >30% report moderate to severe symptoms (frequent to daily interference with activities)
  - Males and those with more severe disability had more severe spasticity symptoms
- Spasticity-related pain complaints very prevalent in persons with MS
- Fine balance between alleviating symptoms and not limiting functional aspects of spasticity
Spasticity in ALS

- Spasticity is common and decreases quality of life in persons with ALS

- Spasticity management in ALS is different from other spinal cord disorders- botulinum toxin injections are relatively contraindicated in progressive neuromuscular disease
Spasticity Management Toolkit

CONSERVATIVE TREATMENT
MEDICATIONS
INJECTIONS
INTRATHECAL PUMP
SURGERY
Conservative Treatment for Spasticity

Stretching
Splinting
Modalities
Stretching for Spasticity Management

- **Why:**
  - Maintains/improves range of motion
  - Reduces the stretch input from the muscle spindle which triggers spasticity
  - Additive benefit of stretching combined with chemodenervation

- **How:**
  - Weightbearing stretches may be superior to non-weightbearing stretches to diminish spasticity
  - Prolonged stretching >1 minute may be more effective
Splinting for Spasticity

• **Why:**
  - Prolonged stretch
  - Diminished sensory input while in the splint
  - Maintains range of motion

• **How:**
  - Serial casting
  - Static splints
  - Dynamic splints
Weightbearing stretches
Modalities for Spasticity

- **Why:**
  - Low risk
  - Provide tools for self-management

- **How:**
  - Vibration
  - Electrical Stimulation
  - Cryotherapy
  - Heat

Caution in neurogenic skin!

Photo courtesy of Vanessa Roberts, OTR/L
Exercise

- Evidence for exercise as a primary or adjunctive treatment for spasticity
  - Land-based
  - Aquatic exercise
  - Functional Electrical Stimulation

- Includes ALS among conditions benefiting from exercise with improved spasticity outcomes
Conservative Interventions: Summary

- Multiple interventions available for spasticity management:
  - Stretching
  - Splinting
  - Exercise
  - Vibration
  - Electrical Stimulation
  - Heat/cold

- Optimal *advanced* spasticity management requires:
  - Coordination of appointments with therapies/"pathway"
  - Communication/feedback with interdisciplinary team
Medications
Medications

Principles
- One of the first interventions
- Global as opposed to local involvement

Considerations
- Adverse effects
- “Pill burden”
Medications

Most commonly used types

- Central acting
  - GABA analogs
  - Alpha 2 adrenergic
- Peripheral acting
Gama-aminobutyric acid (GABA) receptor system

- Chief inhibitory neurotransmitter of the central nervous system
- Two main classes-A and B
- Indirectly inhibits action potential formation in alpha motor neuron
# Medications

## Benzodiazepines
- **Diazepam, Clonazepam**
- **Action**
  - GABA-A receptors
  - Does not directly mimic GABA
- **Dosage - Diazepam**
  - 2-10 mg 3-4 times a day
- **Dosage – Clonazepam**
  - 0.5-1 mg 3 times a day

## Baclofen
- **Action**
  - GABA- B receptors
  - Mimics GABA molecule
- **Dosage**
  - 5-20 mg 4 times a day
Medications

Benzodiazepine cont.
- Adverse effects
  - sedation
  - cognitive impairment
  - dependence
  - withdrawal
  - seizures

Baclofen cont.
- Adverse effects
  - drowsiness
  - fatigue
  - weakness
  - nausea
  - dizziness
  - hallucinations
  - seizures with stoppage of medication
Medications - central acting

Alpha 2 adrenergic receptor system

- Receptors found throughout the body
  - Central - causes sedation and analgesia
  - Peripheral - causes bradycardia, diuresis, and vasoconstriction or dilation

- Mechanism of action
  - Inhibit the release of excitatory amino acids in spinal interneurons
Medications

α2-adrenergic agonists

- **Clonidine**
  - Initial 0.1mg BID
  - Up to 2.4mg total daily

- **Tizanidine**
  - Initial 2-4 mg at bedtime
  - Max dosage of 8mg TID

- **Adverse Effects**
  - dry mouth
  - drowsiness
  - dizziness
Dantrolene

- Decreases the release of calcium ions in skeletal muscle causing weakened contraction

- Dosage
  - Up to 100 mg 3 times a day

- Adverse effects:
  - Drowsiness
  - Diarrhea
  - Malaise
  - Hepatotoxicity
Advanced Spasticity Management

Chemodenervation: Botulinum toxin
Injections

Principles:
- Focal treatment
- Reduce exposure to side effects of systemic medications

Considerations
- Invasive
- Painful
- May require repeated treatments
Botulinum Toxin

Most common question
Botulinum Toxin
Botulinum Toxin

History

- 1895 – bacteria identified by van Ermengem
- 1920 – purified as acid by Sommer
- 1946 – crystalized by Schantz
- 1950s – mechanism of action
- 1960 – Scott researched use of toxin for strabismus and for Oculinum Inc.
- 1988 – Allergan acquires Oculinum
- 1989 – name changes to Botox and first FDA approval
- 2010 – Botulinum toxin approved for use in upper limb spasticity
Botulinum Toxin

Type A

- Botox
  - onabotulinumtoxinA (US - Allergan)
- Dysport
  - abobotulinumtoxinA (Medicis - Ipsen Pharma. - UK)
- Xeomin
  - incobotulinumtoxinA (Merz - Germany)
- Neuronox, Siax
  - Purified Botulinum Toxin Type A Complex (Medytox - S. Korea)
Botulinum Toxin

Type B
- Myobloc, Neurobloc
  - rimabotulinumtoxinB (Solstice-US)
Botulinum Toxin

- Most common treatment for focal spasticity
- Inhibits release of acetylcholine at the neuromuscular junction
- Weakens muscle
- Starts to work in 3-7 days and can last up to 6 months
- Cost – expensive - $500/100 units for BOTOX®
Botulinum Toxin

- Sold as crystalline and diluted at time of use in normal saline
  - Wide variation in dilution practice
    - 2-4 cc / 100 units
  - Proteins are delicate, caution during reconstitution
  - Units of one type botulinum toxin are not convertible to another

- Typically 100-400 Units of BOTOX® Type A per session

- Recommended maximum of BOTOX® is 360 Units/3 months.
Botulinum Toxin

Contraindications
- Prior allergic reaction
- Injection into areas of infection or inflammation
- Pregnancy
- Breast feeding

Caution
- Myasthenia Gravis, ALS
- Medications
  - Aminoglycosides, calcium channel blockers, penicillamine, quinine
Botulinum Toxin

Complications

- dry mouth
- reduced sweating
- dysphagia
- weakness
- death
Botulinum Toxin

Dosing Adjustments
- Voluntary control
- Ashworth score
- Patient weight
- Muscle bulk
Botulinum Toxin

- Recommended dilution of BOTOX® is 100 U/ 2cc Normal Saline
- No more than 50 U or 0.5-1cc volume/site
- 25-30g needle for superficial muscles
- 22 g for deeper muscles
- EMG for localization
Botulinum Toxin

Selected upper extremity conditions treated with BOTOX®

• Flexed elbow:
  ○ Biceps, Brachialis, Brachioradialis
    ▪ 100-200 U divided in 4 sites

• Flexed wrist
  ○ Flexor Carpi Radialis and Ulnaris
    ▪ 12.5-50 Units in each muscle in 1 site

• Clenched fist
  ○ Flexor Digit. Profundus and Superficialis
    ▪ 30-50 U in each muscle in 1 site
Selected lower extremity conditions treated with BOTOX® - not currently approved by FDA

- **Flexed hip**
  - Iliopsoas and Rectus Femoris
    - 100 U in each muscle in 2 sites

- **Flexed knee**
  - Medial and lateral hamstrings
    - 100 U in each muscle in 2-3 sites

- **Plantar flexed foot**
  - Medial/lateral Gastrocnemius and Soleus
    - 100 U in each muscle in 2-4 sites
Advanced Spasticity Management

Chemodenervation: phenol neurolysis
Phenol neurolysis

- Lost art?
- Secret tool?
Phenol - history

- Also known as carbolic acid
- Low doses (<1%) provide topical analgesia
- 1865, Lister employed 5% phenol as an antiseptic for covering surgical wounds
- At high doses, it denatures proteins and functions as a sclerosing agent - 50% phenol used to treat hemorrhoids in early 1900s
Phenol

Long history of use for spasticity and pain:

- 1950: Mandle injected phenol for sympathectomy

- 1955: intrathecal phenol used for intractable cancer pain

- 1959: Nathan, Kelly & Gauthier-Smith injected intrathecal phenol for spasticity

- 1965: Halpern and Meelhuysen described motor point blocks with 5-7% phenol
  - No incremental benefit from 7% over 5%

- 1967: Khalili performed selective nerve blocks with 2-3% phenol
How does phenol work?

- Low dose phenol blocks sensory nerves, possible demyelination

- 4-5% phenol causes inflammation, then destruction and Wallerian degeneration of nerve fibers

- Toxicity: 8.5 g phenol toxic (170 ml of a 5% phenol solution)
  - Typical total injection doses ~3-5 ml of 5% phenol for neurolysis
Phenol- indications

- Localized spasticity

- Large muscle involvement
  - Makes Botulinum toxin less practical due to max dose

- Medications not tolerated/contraindicated
  - Children
  - Abnormal liver function

- Conservative treatment not effective
Phenol- contraindications

• Same as for Botulinum toxin:
  - Local/systemic infection
  - Pregnancy
  - Progressive neuromuscular weakness
  - Caution in causing too much weakness in functional muscles
    - Muscles required for ambulation, posture, etc...
Phenol- benefits

- Cost

- Longer duration of effect (6-9 months vs. 3-4 months from BoNT)
  - Possibly beneficial for patients who live far from the medical center

- Immediate effect- helpful in the acute rehab setting
Phenol- pitfalls

- Time
- Experience
- Repeat injections more technically challenging
- Risk of dystesthesias when used on mixed nerves
  - Reported incidence: 0-32%
  - Presents days-2 weeks following injection
  - Duration: months-year
- Risk of unintended nerve damage/spinal injury (if injected near spine)
- Sudden loss of functional tone
Phenol- procedure basics

Equipment needed:

- EMG machine or peripheral nerve stimulator
- Injectable EMG needle ~22g, 50-75mm
- [IV tubing]
- Phenol 5% aqueous solution
Phenol- summary

- Slightly sharper learning curve than botulinum toxin injections
- Longer duration of effect than botulinum toxin
- Shorter onset of action than botulinum toxin
- Less dosing limitation for larger muscle groups
- Good option for hip adductor spasticity to facilitate hygiene or seating
Advanced Spasticity Management

- Intrathecal Baclofen Pump
Intrathecal Baclofen (ITB)

- **Indication:** Management of severe spasticity not adequately controlled with maximum or tolerable dose of oral agents and/or other techniques

- **Patient selection:** invasive surgery; close follow up; no wounds/infections

- **Trial:** Bolus dose injected into the CSF via lumbar puncture and response is evaluated in clinically supportive setting.
Intrathecal Baclofen (ITB) Trial

- Physiologic evaluation for response
- Pre-infusion physical and functional assessment
- Lumbar puncture
- Infuse 50mcg bolus

- Observe for 4-6 hours
- Post-infusion assessment
- Positive result: desired result
- Negative result: insufficient response
- Repeat trial at 75-100mcg on another day
Intrathecal Baclofen (ITB)

- **Implantation:** Infusion pump is implanted in subcutaneous tissue of abdominal wall with catheter delivering precise, concentrated dose of baclofen to the spinal cord - intrathecal space - directly into the CSF.
Intrathecal Baclofen Pump

- Medication refills are required periodically via percutaneous puncture into access port
Benefits with ITB

Benefits as compared with oral meds:
- High concentration of baclofen – $1/100^{th}$ of oral dose
- Reduce likelihood of negative side effects
- Precise dose adjusted / customized for optimal effect
- Effective in SCI&D: 97% (Penn, 1992) potentially reducing Ashworth Score (Dijkers, 1996)
- May also provide improved pain management, decreased bladder hyperactivity, time saving in ADLs and for caregivers
Risks with ITB

- Risks: overdose, withdrawal or infection possibly related to:
  - Catheter or pump moving or eroding through skin
  - Catheter: disconnection, leak, tear, kink, blockage, dislodgement, migration
  - Pump empty
  - Pump failure
  - Battery depletion
Withdrawal of ITB

- **Symptoms commonly seen:**
  - High fever, sweating
  - Itching without pruritis
  - Worsened spasticity and muscle rigidity
  - Altered mental status – irritability

- **Add caution for:**
  - Patients on high doses
  - Non-verbal
  - In care facility
  - After pump replacement

- **Patient should have supply/prescription on hand**
ITB Clinical Pearls

- ITB catheter is most commonly seated in the lumbar spine region for control of lower extremity spasticity, but can be placed higher

- Patient, caregivers, providers must be educated on withdrawal symptoms and treatment

- Patients/caregivers in remote/rural areas need support

- Pump low volume alarm: most common few days after refill

- Pump low volume alarm: may not be heard/noticed in loud care setting

- Choose concentration of ITB that minimizes refill frequency but no longer than 6 months

- MRI compatible but stops then restarts - needs to be checked at 2 hours
Surgery
Surgery

- **Rhizotomy**
  - Selectively severing parts of nerve roots
  - Mostly done in children

- **Tenotomy**
  - Total or partial severing of a tendon
  - Reserved for severe spasticity
  - No voluntary movement
  - Used to help position a joint in position for seating or better care
Algorithm for Spasticity Management
<table>
<thead>
<tr>
<th>Algorithm – patient considerations</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Focal or diffuse?</td>
</tr>
<tr>
<td>• Upper or lower limbs affected?</td>
</tr>
<tr>
<td>• Results/adverse reactions from prior treatments</td>
</tr>
<tr>
<td>• Patient exam</td>
</tr>
<tr>
<td>• Compliance</td>
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<tr>
<td>• Goals of care</td>
</tr>
<tr>
<td>• Preference</td>
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Algorithm - treatment considerations

- Medications
- Injections
- ITB Pump
- Surgery

Conservative Treatment
Factors Contributing to Spasticity

- Important to minimize / treat contributing factors prior to treatment
  - Infection
  - Wound / pressure ulcer
  - Syringomyelia (syrinx)
  - Changes in level of activity, i.e. bed rest
  - Other noxious stimuli: full bladder/bowels, skin irritation, heat/cold, fracture, tight clothing, binders, discomfort from seating system
Algorithm

- Cannot rely on just one treatment option, but a combination
- Must constantly reassess patient and situation and adjust
STAR ALGORITHM

- Conservative Treatment
- Surgery
- Medications
- Pump
- Injections
Case Examples & Application of the Algorithm

Case 1
Case #1

- 68-year old gentleman with Secondary Progressive MS

- Functionally, he uses a power chair for mobility, and is able to perform stand pivot transfers independently. He has a caregiver available for intermittent assist as needed

- He has gradually noticed increasing difficulty with transfers due to “leg stiffness”
  - His main stated goals are to improve his transfers and maintain independence
Case #1, continued

• Prior treatment:
  
  o baclofen 20mg qid, which he is currently taking
  o He could not tolerate cognitive effects of tizanidine
  o He had physical therapy in the past for range of motion exercises, but doesn’t follow any routine program at this time
  o No splints
Case #1: Physical Exam

- Physical exam reveals a jovial, heavy-set gentleman seated in a powerchair

- Range of motion evaluated- ankle dorsiflexion most limited with knees extended

- Lower extremity strength is 3-4/5 throughout

- Spasticity in the bilateral quadriceps MAS 2, sustained clonus at the ankles
You ask the gentleman to demonstrate a transfer in the exam room.

On observation of the patient’s transfer, he is able to pull up to a squatting position with some difficulty.

Sustained clonus is noted in this position, and the patient braces himself for stability.

Quadriceps spasticity does not appear to interfere with his transfer.
APPLICATION OF ALGORITHM

Conservative Treatment

Surgery

Patient

Medications

Pump

Injections
Patient treatment

- Botulinum toxin injections were performed to the gastrocnemius bilaterally

- The patient is referred to physical therapy to reinforce weightbearing stretches and nighttime splinting
Patient follow-up

- He follows up in two weeks.
  - Improved range of motion at the ankles
  - Improved stability during transfers
  - He is very satisfied with the result of the treatment

- He returns for follow-up at three months and reports effects have begun to wane, therefore he requests repeat injection
Case Examples & Application of the Algorithm

Case 2
Case #2

- 56 y.o. male Vet with C6 ASIA B tetraplegia; h/o syringomyelia, neurogenic bladder/bowel (SPT), spasticity & increased tone in LE bilaterally

- Functionally, he uses a manual chair with power tilt feature for mobility, transfers with ceiling lift with assistance

- Complains of muscle spasms resulting in pain, interference with ADLs & hygiene, increased pressure in seating and heel wounds from friction in lower extremities
Case #2, continued

- Patient’s goal - better manage spasticity to:
  - Minimize pain
  - Decrease effort by him and caregivers for ADLs allowing improved hygiene and skin care
  - Reduce/eliminate increased pressure on boney prominences especially in lower extremities allowing healing of pressure existing pressure ulcers and decrease risk of further ulcers
Case #2, continued

- Prior treatment:
  - baclofen 20mg qid - higher doses resulted in fatigue and only marginal decrease in symptoms

- Physical exam:
  - thin, knees wind swept, manual w/c with power tilt

- Range of motion:
  - diminished due to increased tone & spasticity in hip flexors, hamstrings, adductors, but reducible
STAR ALGORITHM

- Conservative Treatment
- Surgery
- Medications
- Pump
- Injections

Patient
Case #2 - Patient Treatment

- Continue conservative measures
- Continue treatment with oral medications
- Reduce/avoid contributing factors
- Treat heel wounds:
  - osteomyelitis, subtotal calcanectomy with surgical closure
- Scheduled for ITB trial
- Seating optimized but...
  - difficult with prominent ITs even with tilt
  - developed UTI with leaking around catheter
  - developed pressure ulcer from moisture and pressure
Case #2 - Patient Treatment (cont.)

- Continue treatment with conservative treatment and oral medications
- Reduce/avoid contributing factors
- Local wound care, osteomyelitis, flap surgery
- Schedule for ITB trial – trial successful
- Pump implantation and begin ITB therapy
Patient follow-up

- **Post-implant assessment:**
  - Improved range of motion in lower extremities but spasticity still elicited with stretching
  - Proper alignment/positioning in wheelchair enabling seating system to successfully off-load pressure over high risk areas
  - Reports decrease in pain related to spasticity and with more comfortable positioning in chair

- **Follow up assessment: Patient reports:**
  - Reduced caregiver burden
  - Improvement in hygiene and skin care
  - Reduced pain related to spasticity
  - No new skin breakdown
Treat your patients like stars!
Thank you!