Limb Spasticity in Children and Adults

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Disclosures

Dr. Dashtipour has disclosed that he has served as a consultant/independent contractor for AbbVie Inc., Allergan plc, Cynapsus Therapeutics, Inc., Impax Laboratories, Inc., Ipsen Biopharmaceuticals, Inc., Lundbeck, Merz, Inc., Teva Pharmaceuticals, and US WorldMeds LLC. He has also served as a Speaker’s Bureau participant for AbbVie Inc., Allergan plc, Acadia Pharmaceuticals, Inc., Impax Laboratories, Inc., Ipsen Biopharmaceuticals, Inc., Lundbeck, Merz, Inc., Teva Pharmaceuticals, UCB, and US WorldMeds LLC.

Dr. Przekop has nothing to disclose.

This lecture will discuss off label uses of the botulinum neurotoxins.
Goals and Objectives

- To describe the clinical aspects of the diagnosis of spasticity in adults and children.
- To provide the clinician with available options for treatment in patients with spasticity.
- Allow clinicians to determine the process of determining candidates for treatment of chemodenervation.
- Interpret the status of current research in spasticity.
Case Vignette

- Embolic Stroke
  - Hemiparesis, arm more than leg
  - In rehab, difficulty with vision and with daily living skills
  - Family would like to maximize independence

![Brain Scan Image]
Definition of Spasticity

- Motor disorder characterized by a velocity dependent increase in tonic stretch reflexes (muscle tone) with exaggerated stretch reflex.
- Muscle tone: sensation of resistance that is encountered as a joint that is passively moved through a range of motion.

Spasticity

- Stroke
- Multiple sclerosis
- Spinal cord injuries
- Traumatic brain injury
- Cerebral Palsy
- Review of 24 studies with prevalence in 28-38% of stroke, 41-66% in patients with MS and 13 in TBI

<table>
<thead>
<tr>
<th>Score</th>
<th>Description</th>
</tr>
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<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 catch and release or by minimal resistance at the end of the range of motion when the affected part is moved in flexion or extension</td>
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<tr>
<td>1+</td>
<td>Slight increase in muscle tone, manifested by a catch, followed by minimal resistance throughout the remainder of the range of motion (ROM)</td>
</tr>
<tr>
<td>2</td>
<td>Marked increase in muscle tone through most of the ROM, but affected part easily moved</td>
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<tr>
<td>3</td>
<td>Considerable increase in muscle tone, passive movement difficult</td>
</tr>
<tr>
<td>4</td>
<td>Affected part rigid in flexion or extension</td>
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Upper Motor Neuron Syndrome

- Positive symptoms
  - Spasticity
  - Clonus
  - Flexor/extensor spasms
  - Hyper-reflexia

- Negative symptoms
  - Weakness
  - Paralysis
  - Fatigability
  - Weakness of dexterity
Stroke/Spasticity

- 20 to 40% develop spasticity
- Usually three months post stroke
- Co-contraction, muscle shortening and loss of volitional control
- Difficulties with hygiene (axilla, palms, groin) self care, inability to perform activities of daily living.
- Difficulties with assisting caregivers, for example with transfers: sitting to laying etc.
- Improvement in body image and esteem.
Impact of Spasticity

- Pain
- Contracture
- Functional limitations
  - Upper limb: hygiene, dressing
  - Lower limb: hygiene, transfers, seating, walking
- Fatigue
- Nutritional compromise
- Poor self-image due to abnormal limb posture
- Increased risk of falls
- Prolonged immobility
  - Skin breakdown
  - Infection

Pediatric Spasticity Overview

Treatment Options

- Physical/Occupational therapy
- Bracing/Splinting
- Medications
  - Baclofen
  - Tizanidine
  - Benzodiazepenes
- Exercises
  - Improve strength and activity (15 RCT)
- Chemical denervation
  - Botulinum toxin
Oral Medications for Spasticity

- GABA systems
  - Benzodiazepines
  - Baclofen
  - Gabapentin

- Alpha adrenergic systems
  - Tizanidine
Adjunctive Treatment

- Neuromuscular paralyzing agent that is produced by anaerobic bacterium
- Inhibits the release of acetylcholine.
- Toxins are taken up by endocytosis at the cholinergic nerve terminals where they block the releases of synaptic vesicles.
- Dose dependent chemical denervation resulting in reduced muscle activity
- Reversed after several months when nerves generate new neuromuscular junction by terminal sprouting
Chemodenervation

- Actions of Botulinum toxin
  - Inhibition of spastic co-contraction
  - Dystonia
  - Contribution to ease and stretch by lengthening the muscle
  - Help to increase the antagonist torque
Untoward Side Effects

- Immune resistance
- Diffusion of toxin can lead to unwanted inhibition of transmission at neighboring nerve endings
- Atrophy, similar to anatomic denervation
Common Clinical Patterns

The Flexed Wrist

The Pronated Forearm

The Flexed Elbow

The Thumb-in-Palm Deformity

Upper Extremities

Pectoralis

Biceps brachi, medial and lateral heads
Upper Extremities: Thumb Adduction

Adductors/ First dorsal Interossei
Upper Extremities: Flexors Excessive Flexors

- Flexor carpi ulnaris
- Flexor digitorum superficialis
- Flexor carpi radialis
- Pronator teres
Spasticity /Stroke

- Botulinum, AAN recommendations: 11 Class 1 trials. Functional improvement, one study showed improvement in disability score.
- Randomized clinical trials, One study with 96 patients and another with 333 adults, showed a reduction in modified Ashworth score but no improvement in functions.

Side Effects

Systemic side effects
- Higher dose
- Bulbar compromise
- Diffusion of toxin can lead to unwanted inhibition of transmission at neighboring nerve endings

Local side effects
- Anxiety
- Pain
- Weakness
- Calf atrophy
Management Goals

- Decrease tone and improve range of motion
- Enhance fit of orthoses
- Decrease contracture
- Facilitate rehabilitation
- Enhance appearance
- Decrease caregiver burden
- Improve functioning
  - Activities of daily living, Mobility, Sleep, Dressing
Management Team

- Physical/Occupational therapy
  - Maintain muscle strength, improve body symmetry and functional activity
- Splints/Orthoses
  - Improve gait, balance, improve posture and alignment
- Casting
  - Helps stretch muscles and facilitate growth, improve muscle tone, gait and passive range of motion
Spasticity in Infants and Children

- Cerebral palsy
  - Static lesion producing motor and movement disorder with an insult occurring early in life.
  - Learning difficulties
  - Hearing/ vision abnormalities
  - Spasticity
    - Gait
    - Pain
    - Quality of life
Case Vignette

- 2 year old born prematurely at 30 weeks.
- Toe walking since fourteen months of age.
- MRI evidence of periventricular leukomalacia
- Exam: spasticity: lower extremities > upper ext
Cerebral Palsy

- 2 in 1000 live births
- Most common cause of physical disability
- Spastic motor type: 80%
- Other types: Dystonia
Differential Diagnosis of Causes of Spasticity in Infants and Children

- Cerebral palsy
- Dopa-responsive dystonia
- Genetic diseases
  - Rett’s syndrome
- Neurodegenerative disease
  - Batten’s disease
  - Huntington’s disease
Dopa Responsive Dystonia

- Dystonia
- Parkinsonism with diurnal fluctuations
- Improves with sleep /or res
- Marked responsiveness to L-dopa
- Mimics CP
- May have developmental delay
- Diagnosis: Gene testing.
Dopa Responsive Dystonia

- Acutely responsive to carbidopa levodopa.
- Diagnosis: Gene testing: whole exome, targeted genes, spinal taps.
- Clinical presentation
  - Toe walkers, Parkinsonian features, Diurnal variations
  - CP-Like
Case Vignette

What should you do next?

- Passive stretching
- Facilitation of motion and movement
- Splinting/Casting
- Facilitation of motion and movement
- Medications
- Therapies
Lower Extremity Measurement of Function

- Ashworth scale
- Pediatric evaluation of disability index (PEDI)
- Gross motor function measure (GMFM)
- Goal attainment scale
- Videos
- Canadian occupational measurement
Outcome Measures

- Physical exam
- Grip and pinch strength
- Tardieu scale
- Muscle tone (Ashworth, Modified Ashworth)
- QUEST
Lower Extremity

- Medial and Lateral gastrocsoleus muscles
- Adductors
- Gastrocsoleus
- Posterior tibialis muscle
Lower Extremity

- Excessive knee flexion throughout gait cycle
- Equinus during gait cycle Failure to dorsiflex beyond neutral
- Non ambulatory: prevent skin breakdown
- Children/ Infants: pain; aid with care; transfers
Lower Extremity

- Lateral and Medial Gastrocsoleus
  - Equinovarus and Equinovalgus
- Adductors
  - Scissoring (adductor)
- Hamstrings
  - Knee straightening and Standing ability
  - Crouch gait
Common Clinical Patterns: Lower Limbs
- Spastic Hemiplegia
- Crouch Gait
- Apparent Equinus
- Spastic Diplegia
Lower Extremities
Surgical Options

- Focal surgery, with release/lengthening of tendon
- Intrathecal Baclofen
  - Spastic diplegia
  - Anoxic brain injury
  - Drowning.
- Dorsal Rhizotomy
Goals of Treatment

Russman BS, Tilton A, and Gormley ME Jr.

- Maximize function,
  - Minimize the development of secondary problems such as contracture, thereby delaying or obviating the need for surgical intervention
  - Enhances run/balance.
  - Enhance ADL’s to minimize caregiver stress

Pediatric Spasticity

Outcome Measures

- Muscle tone
  - Ashworth Scale
  - Modified Ashworth Scale
- Muscle reaction at different velocities of stretch
  - Tardieu Scale
- Range of motion
- Muscle strength
- Electrophysiology
  - M-response
  - H-reflex
- Disability / functioning
  - Pediatric Evaluation Disability Inventory
  - Functional Independence Measure for Children
- Overall motor function
  - Gross Motor Function Measure
- Gait
  - Physician Rating Scale
  - 3-dimensional motion analysis
- Caregiver assessment
Conclusions

- Clinician is able to diagnose and treat adults and children with diagnosis of cerebral palsy
- Rational therapy in the management of children with cerebral palsy
- Formulate a plan for long term care