PATHOPHYSIOLOGY of SPASTICITY

Patricia Tan, MD, FAAPM & R
Assistant Professor
Department of Rehabilitation and Regenerative Medicine
Department of Pediatrics
Columbia University Medical Center

Medical Director
Multidisciplinary Spasticity Center
Morgan Stanley Children’s Hospital of New York
New York Presbyterian
LEARNING OBJECTIVES

• Define and differentiate spasticity from other neurological conditions associated with increased tone.
• Delineate the pathophysiology of spasticity.
• Discuss the various presentations and outline functional outcome measures.
• Understand the natural history and progression of spasticity including the various etiologies and sequelae.
• Define various treatment options available.
• Discuss the indications, technique and outcome of injectable treatment of spasticity.
Epidemiology of Spasticity

• Spasticity affects more than 12 million worldwide

• Specific to type and severity of CNS injury
  - 65-90 % of Cerebral Palsy
  - 65-78 % of Spinal Cord Injury
  - 5-56 % of Cerebrovascular Accidents

• Prevalence vary depending on etiology
  – 80% affected with UE Impairment 6 months after CVA

  Lang, 2009, Archives

NewYork-Presbyterian
The University Hospital of Columbia and Cornell

Columbia University Medical Center
Spasticity

• Motor disorder
  – Occurs in response to passive muscle stretch
  – **Velocity-dependent** increase in muscle tone
  – Exaggerated tendon jerks
  – Resulting from hyper-excitability of muscle stretch reflex
Etiology

• Injury to the Central Nervous System (CNS)
  – Cerebral Palsy
  – Traumatic Brain Injury
  – Cerebrovascular Accidents
  – Spinal Cord Injury
  – Multiple Sclerosis

• Neural reorganization

• Adaptive pathologic changes in peripheral muscle control
Etiology

- Prolonged immobility
- Muscle disuse
- Heightened afferent muscle stretch
- Efferent contraction signals
- Diminished inhibition of muscle contraction
- Hyperactive reflex arc
Patho-anatomy depends on the site of CNS injury

Damage to an upper motor neuron

- Loss of inhibitory control by descending corticospinal tracts
- Increased dependence on rudimentary brainstem-mediated descending motor tracts
- Maladaptive branching of intact corticospinal fibers
- Increased sensitivity of stretch-activated spindles
- Pathological branching of spinal interneurons
  - Results in hyper excitability of alpha motor neuron
  - Innervating the involved muscle
Disease Progression

- **New onset/acute**
  - Flaccid
  - Neurogenic shock
- **Subacute**
  - Muscle tightening or cramping
- **Chronic**
  - Bony deformity
    - Tibial torsion
    - Hip dysplasia
Disease Progression

- Chronic
  - Remodeling of soft tissue
  - Muscle and soft tissue contractures
    - Muscle atrophy
    - Loss of sarcomeres
    - Accumulation of connective tissue
    - Increase in muscle responsiveness
  - Range of motion limitation
  - Postural and functional abnormalities
Secondary Conditions

- Pressure ulcer
- Contractures
- Poor hygiene
- Joint subluxation
- Impaired mobility
- Poor balance
- Loss of selective motor control: strongest predictor of gross motor capacity in CP
## UMN Syndrome

<table>
<thead>
<tr>
<th>Positive signs: abnormalities that lead to involuntarily increased muscle activity or movement patterns</th>
<th>Negative signs: reflect insufficient muscle activity or control of muscle activity interfering with function</th>
</tr>
</thead>
<tbody>
<tr>
<td>Spastic hypertonia (velocity dependent)</td>
<td>Motor weakness</td>
</tr>
<tr>
<td>Spastic dystonia</td>
<td>Muscle fatigue</td>
</tr>
<tr>
<td>Spastic co-contraction of agonist and antagonist</td>
<td>Loss of selective motor control</td>
</tr>
<tr>
<td>Hyperreflexia (due to hyper excitability of stretch reflex)</td>
<td>Impaired fine motor function</td>
</tr>
<tr>
<td>Clonus</td>
<td>Slow effortful voluntary movements</td>
</tr>
<tr>
<td>Extensor plantar responses</td>
<td></td>
</tr>
</tbody>
</table>

**NewYork-Presbyterian**

**Columbia University Medical Center**

*Discover. Educate. Care. Lead.*
Pathophysiology of Spasticity

UMN lesion

Muscle overactivity

Dynamic Spasm Co-contracture Clonus Associated reaction Flexor withdrawal

Weakness

Immobilization of short muscle length

Biomechanical changes reduced compliance contracture

Hypertonia Reduced ROM

Abnormal posture

Impaired function

### UMN Patterns Static and Dynamic Forces

#### UPPER LIMB

<table>
<thead>
<tr>
<th>Muscles/Patterns</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pectoralis major</td>
<td>The Adducted/Internally Rotated Shoulder</td>
</tr>
<tr>
<td>Brachialis (elbow flexion)</td>
<td>The Flexed Wrist</td>
</tr>
<tr>
<td>Brachioradialis</td>
<td>The Pronated Forearm</td>
</tr>
<tr>
<td>Biceps</td>
<td>The Clinched Fist</td>
</tr>
<tr>
<td>Pronator teres</td>
<td>The Flexed Elbow</td>
</tr>
<tr>
<td>Flexor carpi radialis</td>
<td>The Thumb-in-Palm Deformity</td>
</tr>
<tr>
<td>Flexor carpi ulnaris</td>
<td></td>
</tr>
<tr>
<td>Flexor digitorum superficialis</td>
<td></td>
</tr>
<tr>
<td>Flexor digitorum profundus</td>
<td></td>
</tr>
<tr>
<td>Adductor pollicis</td>
<td></td>
</tr>
<tr>
<td>Flexor pollicis longus</td>
<td></td>
</tr>
</tbody>
</table>
Spasticity affecting lower extremities

- Adductors magnus
- Semimembranosus
- Semitendinosus
- Biceps femoris
- Rectus femoris
- Extensor hallucis longus
- Posterior tibialis
- Gastrocnemius
- Soleus
Essential of Assessment

- History
  - Severity
  - Location
  - Time of day
  - Aggravating/alleviating factors
  - Interference with function or safety
  - Patterns of overflow
  - Possible benefits (assisting for transfers)
Examination: Clinical Measures of Spasticity

- Range of motion of joints
- Clonus
- Deep tendon reflexes
- Intensity: Modified Ashworth scale
- Associated non-spastic motor abnormalities: dystonia, tremors
- Functional status: GMFCS (Gross Motor Functional Classification Scale)
## Modified Ashworth Scale

<table>
<thead>
<tr>
<th>Score</th>
<th>Description</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 (catch and release feeling)</td>
</tr>
<tr>
<td>1+</td>
<td>Slight increase in muscle tone with minimal resistance throughout remainder of movement</td>
</tr>
<tr>
<td>2</td>
<td>Marked increase in muscle tone but easily moved</td>
</tr>
<tr>
<td>3</td>
<td>Increased muscle tone with difficult passive movement</td>
</tr>
<tr>
<td>4</td>
<td>Rigid without movement</td>
</tr>
</tbody>
</table>
GMFCS E & R
Descriptors and Illustrations for Children between their 6th and 12th birthday

**GMFCS Level I**
Children walk at home, school, outdoors and in the community. They can climb stairs without the use of a railing. Children perform gross motor skills such as running and jumping, but speed, balance and coordination are limited.

**GMFCS Level II**
Children walk in most settings and climb stairs holding onto a railing. They may experience difficulty walking long distances and balancing on uneven terrain, inclines, in crowded areas or confined spaces. Children may walk with physical assistance, a hand-held mobility device or use wheeled mobility over long distances. Children have only minimal ability to perform gross motor skills such as running and jumping.

**GMFCS Level III**
Children walking using a hand-held mobility device in most indoor settings. They may climb stairs holding onto a railing with supervision or assistance. Children use wheeled mobility when traveling long distances and may self-propel for shorter distances.

**GMFCS Level IV**
Children use methods of mobility that require physical assistance or powered mobility in most settings. They may walk for short distances at home with physical assistance or use powered mobility or a body support walker when positioned. At school, outdoors and in the community children are transported in a manual wheelchair or use powered mobility.

**GMFCS Level V**
Children are transported in a manual wheelchair in all settings. Children are limited in their ability to maintain antigravity head and trunk postures and control leg and arm movements.
Functional Assessment

- Bed mobility
- Transfers
- Ambulation
  - Assistive devices
  - Orthotics
- Balance
  - Static
  - Dynamic
- Endurance
- Activities of daily living
Patient/Caregiver Report

- Quality of life
- Global outcome measures
- Satisfaction/preference
- Participation impairments
- Dependence
- Functional status
  - RIC Care and Comfort Caregiver Questionnaire
  - Functional independence Measure (WeeFIM)
Rehabilitation Management and Treatments

• The goal of spasticity management is to temper abnormal tone while improving function
• No universal guidelines
• Stepped approach
  – Physical /occupational therapy
  – Self management techniques
  – Pharmacological
  – Procedural interventions
Management interventions

- Physical and occupational therapy
- Orthopedic surgery
- Neurosurgery
- Medication: Oral Intrathecal Neuromuscular
Treatment of focal spasticity

Neurolytics
- Phenol 3%-6%
- Dehydrated alcohol 35-60%

Chemodenervation
- Onabotulinumtoxin A (Botox)
- Rimabotulinumtoxin B (Myobloc)
- Abobotulinumtoxin A (Dysport)
- Incobotulinumtoxin A (Xeomen)
Alcohol 45%

• Denatures protein; tissue necrosis; does not diffuse
• Used to reduce focal increase in tone; larger muscles
• **Duration of effects:** 6-12 months
• Adverse effects:
  – pain during injection
  – dysesthesia
  – excessive motor weakness
  – skin slough
  – convulsion
Botulinum Neurotoxin Mechanism of Action: binds to presynaptic cholinergic nerves, blocks release of neurotransmitter Ach by blocking synaptosomal associated protein SNAP-25
Chemodenervation

• **Flexibility and strengthening effect**
• Improves balance between spastic agonist and weakened antagonist
• Works with therapies (PT/OT) and bracing/splint
• Rare adverse effects
  - Pain during injection
  - Infection
  - Bleeding
  - Rash
  - Flu like symptoms
  - Allergic reaction
  - Excessive weakness and fatigue
**Single event multilevel chemoneurolysis**

<table>
<thead>
<tr>
<th>Muscles injected</th>
<th>Dose range (IU/Kg of bw)</th>
<th>Number of sites</th>
</tr>
</thead>
<tbody>
<tr>
<td>Biceps</td>
<td>2</td>
<td>2–3</td>
</tr>
<tr>
<td>Pronator teres</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Flexor carpi radialis</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Flexor carpi ulnaris</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Flexor digitorum superficialis</td>
<td>2</td>
<td>1–2</td>
</tr>
<tr>
<td>Flexor digitorum profundus</td>
<td>2</td>
<td>1–2</td>
</tr>
<tr>
<td>Flexor pollicis longus</td>
<td>0.5–1</td>
<td>1</td>
</tr>
<tr>
<td>Adductor pollicis</td>
<td>0.5–1</td>
<td>1</td>
</tr>
</tbody>
</table>
Single Event Multilevel Chemoneurolysis

Medial head of gastrocnemius
Lateral head of gastrocnemius
Soleus
Tibialis posterior
Guidance Techniques

– Optimizing outcomes of procedure
– Reducing BoNT dose required for treatment efficacy
– Minimizing potential risks and adverse events
  • Palpation, surface anatomy
  • **Electrical stimulation (E-stim)**
  • Image localization via **ultrasound**
Pediatric modified evidence-based treatment options

Physical and Occupational Therapy

- Stretching/strengthening
- Orthotics
- **Serial casting**
- Biofeedback
- Constraint-induced movement therapy
- Hippotherapy
- Aquatic therapy
Patterns of spasticity: all impact quality of life

- Mobility
- Hygiene
- Self care
- Sleeping pattern
- Disfigurement
- Self esteem
- Affect and mood
- Sexual function

Patterns of spasticity:
- General: TBI, MS
- Regional: SCI
- Focal: CVA
- Cerebral Palsy
Conclusion/Future

- Spasticity is a type of “muscle over activity.”
- Leads to immobilization, muscle shortening, contracture, disuse, functional decline.
- Management is dependent on the presentation and goals of treatment.
- Comprehensive multidisciplinary approach and coordination of care is most important.
- More research is needed to define criteria for therapies, follow the effect of treatment in order to make more definitive recommendations.
- Cutting edge emerging and unique concepts: robotic rehabilitation
Thank You