Spasticity in the Medically Complex Child

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Complex Care Curriculum

Spasticity
Spasticity

Describe the signs, symptoms and complications of spasticity

Identify the patient with spasticity that is not optimally managed

Summarize the different pharmacologic and non-pharmacologic treatment modalities

Plan a basic approach to managing spasticity in a patient

Identify the role of the consultant (PM&R) in helping with management

Identify the patient with baclofen withdrawal and initiate management
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Spasticity
Spasticity = Motor Disorder

Velocity Dependent
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Spasticity

Learning Objectives
Definition
Etiology & Pathophysiology
Impact of Spasticity & Outcome Measures
Exam Features
Treatment Plan
Treatment Options
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Selective Rhizotomy
Chemodenervation
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Velocity dependent
Stretch dependent
UMN signs
Muscle over-activity
Spasticity

Upper Motor Neuron Syndrome

Positive Symptoms
- Spasticity
- Clonus
- Flexor/extensor spasm
- Hyper-reflexia

Negative Symptoms
- Weakness
- ↓ dexterity
- Paralysis
- Fatigability
- ↓ movement

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#### Brain Injury

- traumatic brain injury, cerebral palsy, stroke, bacterial meningitis, encephalitis, tumor, MS

#### Spinal Cord Injury

- tumor, infection, trauma, MS

#### Neuromuscular

- ALS, Friederich ataxia

#### Genetic disorders and degenerative diseases

- Sjogren-Larsson syndrome, Tay-Sachs disease, and Rett syndrome
Spasticity

Cerebral Palsy:

- Disorders of movement and posture causing activity limitations
- Non-progressive disturbances of developing brain

Underlying lesion is static, but musculoskeletal pathology is progressive
Musculoskeletal Progression in Cerebral Palsy

Static
CNS Lesion

Upper Motor Neuron Syndrome
Spasticity and Weakness
Failure of longitudinal muscle growth
Fixed contracture
Bony torsion
Joint instability
Joint dislocation
Joint and bone degenerative changes

Progressive
Musculoskeletal deformity

Must repeatedly screen children for joint dislocation as they grow
Spasticity
Spasticity

- Motor Control
- Bowel/Bladder
- Skeletal & Skin
- Self Esteem

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Muscle Tone
- Ashworth Scale
- Modified Ashworth Scale

Muscle reaction at different velocity of stretch
- Tardieu Scale

Range of Motion
Muscle Strength
Disability/Function
Overall Motor Function
# Ashworth Scale

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>No increase in tone</td>
</tr>
<tr>
<td>1</td>
<td>Slight increase in tone giving a 'catch' when the limb is moved in the flexion or extension</td>
</tr>
<tr>
<td>2</td>
<td>More marked increase in tone, but limb is easily flexed</td>
</tr>
<tr>
<td>3</td>
<td>Considerable increase in tone, passive movement difficult</td>
</tr>
<tr>
<td>4</td>
<td>Limb rigid in flexion or extension</td>
</tr>
</tbody>
</table>
# Modified Ashworth Scale

<table>
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<tr>
<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>No increase in tone</td>
</tr>
<tr>
<td>1</td>
<td>Slight increase in tone - a catch and release at the end of the range of motion</td>
</tr>
<tr>
<td>1+</td>
<td>Slight increase in tone - catch, followed by minimal resistance in remainder of range</td>
</tr>
<tr>
<td>2</td>
<td>More marked increase in tone through most of range</td>
</tr>
<tr>
<td>3</td>
<td>Considerable increase in tone, passive movement difficult</td>
</tr>
<tr>
<td>4</td>
<td>Affected parts rigid in flexion or extension</td>
</tr>
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### Selective Rhizotomy

### Chemodenervation

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## Tardieu Scale

| V1: As slow as possible (minimizing stretch reflex)* |
| V2: Speed of the limb segment falling under gravity** |
| V3: As fast as possible (faster than the rate of the natural drop of the limb segment under gravity)** |

*V1 measures passive range of motion (PROM)  
** V2 and V3 used to rate spasticity
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Spasticity

Observation of function
Identify movement disorder
Assess sensibility
Determine active and passive ROM
Evaluate spasticity, strength, and reflexes
Evaluate posture, gait, and balance
Spasticity
Spasticity

Treatment Plan ↔ Patient Goals

Impact Patient Function?

Impact Daily Care?
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**Spasticity**

- **Learning Objectives**
  - Decrease Tone
  - Increase range of motion
  - Improve fit and use of orthoses
  - Improve position for care
  - Decrease contractures
  - Delay surgery
  - Decrease caregiver burden
  - Improve function

- **Definition**

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**Treatment Factors**

- Drug formulation
- Drug Frequency
- Drug delivery
- Side effects
- Cost

**Patient Factors**

- Patient and family motivation
- Availability of services and resources
- Compliance

**Disease Factors**

- Generalized vs. Focal
- Comorbidities
- Chronicity
- Prognosis

**Patient**

**Disease**

**Treatment**
Spasticity

Evaluate Patient: Does spasticity present a problem?

- Yes
  - Patient and Caregiver objectives
  - Functional objectives
  - Technical objectives
  - Initiate comprehensive spasticity management program

- No
  - No treatment
  - Continual re-assess
Spasticity Management Team

- Patient
- Caregiver
- Pediatrician
- Physiatrist (PMR)
- Neurologist
- Orthopedic surgeon
- Neurosurgeon
- Therapist: PT, OT, Speech
- Orthotist and Durable Medical Equipment provider
- Other: Social Work, Dietician, Education specialist

As children age, they may be more likely to need orthoses and surgery.
# Spasticity

### What is the Role of the Physiatrist?

- Function at home and in the community
- Orthoses and therapeutic equipment
- Altered muscle tone
- Facilitating the ability of the child and family to set functional goals

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Oral medications

Chemo-denervation

Selective dorsal rhizotomy

Intrathecal baclofen

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# Spasticity

## Medication | Mechanism of Action | Most Common Side Effects
--- | --- | ---
Baclofen | - Spinal cord  
- GABA B agonist | - Lowers seizure threshold
Benzodiazepines: Diazepam, clonazepam | - GABA A agonist  
- Spinal cord, brain | - Drooling
Tizanidine (Zanaflex) | - Alpha-2 agonist  
- Spinal cord, brain | - Hypotension  
- Hepatotoxicity
Dantrolene | - Calcium channel blocker  
- Muscle | - Hepatotoxicity
Trihexyphenidyl (Artane) | - Anticholinergic  
- Central muscarinic receptors | - Dry mouth
# Spasticity

## Medications

<table>
<thead>
<tr>
<th>Medication</th>
<th>Initial Dose</th>
<th>Maximum per day</th>
<th>Doses/Day</th>
</tr>
</thead>
<tbody>
<tr>
<td>Baclofen</td>
<td>2.5-5 mg x 2-3/day 0.6 mg/kg/d tid for &lt;12 mo</td>
<td>40-90 mg (age dependant)</td>
<td>3-4</td>
</tr>
<tr>
<td>Diazepam</td>
<td>0.12-0.8mg/kg</td>
<td></td>
<td>3-4</td>
</tr>
<tr>
<td>Clonazepam</td>
<td>0.01-0.03 mg/kg/day</td>
<td>0.1-0.2 mg/kg/d</td>
<td>2-3</td>
</tr>
<tr>
<td>Tizanidine</td>
<td>2 mg/day</td>
<td>36 mg/day</td>
<td>2-3</td>
</tr>
<tr>
<td>Dantrolene</td>
<td>25 mg/day (0.5mg/kg daily)</td>
<td>12 mg/kg/day or 400 mg/day</td>
<td>2-4</td>
</tr>
<tr>
<td>Artane</td>
<td>1 mg/day</td>
<td>15 mg/day</td>
<td>2</td>
</tr>
</tbody>
</table>

## Treatment Options
- Selective Rhizotomy
- Chemodenervation

## Impact of Spasticity & Outcome Measures

## Etiology & Pathophysiology

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Sedation is one of the common side effects with all oral medications.

To minimize potential sedation, always start low and titrate up slow.

Abrupt withdrawal can result in seizures.

If NPO, can use IV Diazepam.
First Line: Oral Baclofen

AVOID MEDICATION ERROR

✓ Concentration is variable
✓ 1 ml ≠ 1 mg

When prescribing oral baclofen, the dose MUST be verified to avoid fatal medication errors!
Spasticity
Spasticity

Intrathecal baclofen: 50x response of oral baclofen
Who is a candidate?

Severe, generalized tone

Not successfully managed with oral medications

Improvement with test dose of intrathecal baclofen given via lumbar puncture
Spasticity

**PROS:**
- GABA inhibition without side effects in the brain
- Programmable to set best dose

**CONS:**
- Complications of device
- High maintenance of pump

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**Mechanical**
- CSF leak
- Catheter malfunction
- Infection

**Medical**
- Overdose
- Withdrawal
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Baclofen withdrawal

**Early signs:**
- Pruritis
- Dysphoria
- Irritability
- Spasticity
- Tachycardia
- Fever
- Hypertension
- Respiratory Distress

**Late signs:**
- Hyperthermia
- Seizures
- Rhabdomyolysis
- DIC
- Altered Mental Status
- Psychomotor agitation
- Respiratory Distress
- Multisystem Organ Failure

Death

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Troubleshooting Pump Failure

1. Medications
2. Radiographs
3. Spiral CT

Call Neurosurgeon!
Spasticity

What is Selective Rhizotomy?

NERVES

Isolated

Targeted

Destroyed

Improved Spasticity
Spasticity

Who is a Good Candidate?

Ages between 4 and 7 years
“Pure” spasticity
Ambulatory
Absence of fixed contractures
Cooperation with intensive therapy
## Spasticity

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# Spasticity

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### Definition

-Botulinum Toxin Type A
-Phenol

### Mechanism of Action

- **Botulinum Toxin Type A**: Selective motor denervation at the NM junction (presynaptic block of ACh release)
- **Phenol**: Non-selective chemical neurolysis in injected nerve

### Onset

- **Botulinum Toxin Type A**: 24-72 hours
- **Phenol**: < 1 hour

### Duration

- **Botulinum Toxin Type A**: 6-12 weeks (3-6 months functional outcome)
- **Phenol**: 2-36 months

### Side Effects

- **Botulinum Toxin Type A**: Weakness
- **Phenol**: Dysesthesia, skin necrosis

### Pros

- **Botulinum Toxin Type A**: Easy to inject, Limited dose of toxin
- **Phenol**: Low cost

### Cons

- **Botulinum Toxin Type A**: Cost
- **Phenol**: Difficult to inject

## Summary

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Spasticity

**Advantages**
- Temporary*
- Minimally invasive
- Increase ROM
- Learn normal movement patterns
- Safely repeated
- Mimic surgical outcome
- Allows age-selective timing of surgery

**Disadvantages**
- Temporary*
- May decrease functional tone
- Total body dose of toxin limited (BTX-A)
- Cost (BTX-A)
Who is a Good Candidate?

- Focal increase in muscle tone
- Absence of fixed contractures
- Absence of bony/joint problems
- No underlying bleeding disorder
Spasticity

**Early Years**
- May allow postponement, simplification, avoidance of surgery

**Later Years**
- May provide pain relief, improved ease of care, functional goals
Spasticity

Spasticity = 1 component of the UMN syndrome

Spasticity = velocity dependent increased resistance to passive stretch

Underlying lesion is static, but musculoskeletal pathology is progressive

Treatment plan is not “one-size fits all”, but should be customized to patient’s goals

Most spasticity medications CANNOT be stopped abruptly. Should be weaned to avoid seizures, or switched to IV if patient is NPO.
Spasticity


