Tone Management in Cerebral Palsy

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Disclosures

• I am involved in a Dysport sponsored research study
Goals

• Describe tone abnormalities seen in children with cerebral palsy
• Discuss indications for treatment of increased tone
• Review specific therapies for increased tone in cerebral palsy
Case 1

• 8 yo boy with mixed quadriplegic CP
  – Difficulty sleeping at night
  – Hard to get him into his wheel chair because of arching
  – Difficulty with dressing because of tightness
Case 2

• 8 yo girl with dyskinetic cerebral palsy
  – Non-ambulatory
  – Some tightness in the morning causes some discomfort
  – She has limited arm use due to abnormal movements
Case 3

• 10 yo girl with left hemiplegic CP
  – Walks on her left toe
  – Occasional falls
  – Some pain at the end of the day
  – Does not tolerate AFO brace
Definition of Cerebral Palsy

– Permanent
– Disorder of movement and posture
– Causes activity limitation
– Caused by nonprogressive problem in the developing brain
Hypertonia

- **Tone**: resistance to passive stretch while the patient is attempting to maintain a relaxed state of muscle activity
  - excludes resistance due to joint, ligament, or skeletal properties

- **Hypertonia**: abnormally increased resistance to externally imposed movement about a joint.
CP: motor subtypes

- Spastic
- Dyskinetic
  - Dystonic
  - Choreoathetoid
- Mixed
- Hypotonic
- Ataxic
Spasticity: definition

- A motor disorder characterized by a velocity-dependent increase in tone
- Commonly associated with exaggerated deep tendon reflexes, resulting from hyperexcitability of the stretch reflex
Measuring Spasticity: Tardieu

Tardieu angle = R2-R1

R2=R1: no tardieu angle, no spasticity
R2 abnormal? Contracture
Contracture

*Permanent tightening of non-bony tissues, such as muscles, tendons, ligaments, or skin.*

- Does not respond to tone management therapies
- Other techniques may help, e.g. serial casting, surgery
Dystonia: definition

Involuntary sustained or intermittent muscle contractions causing twisting and repetitive movements, abnormal postures, or both.

• Voluntary movement overflow is characteristic
• May have co-contraction of agonist and antagonist muscles
• Tone often normal at rest; abnormal postures triggered by movement
Spasticity you feel

Dystonia you see

-Barry Russman, MD
Why Treat Increased Tone?

• Pain – spasms, hip dislocation, pain with movement
• Function – gait/mobility, use of hand, ability to communicate, play sports, independence, etc.
• Other quality of life – dressing, bathing, diapering, maintaining hygiene
• Preventative – contractures, surgery
• Cosmetic
Tone Management Options

Establish care with orthopedic provider for monitoring and management of musculoskeletal complications (scoliosis, hip dislocation, etc.)

1. Serial casting/orthoses
2. Oral Medication
3. Chemodenervation: Phenol, Botulinum toxin injections
4. Selective Dorsal Rhizotomy (spasticity)
5. Intrathecal baclofen (ITB)
6. Deep Brain Stimulation (dystonia)
Oral Medication: spasticity

- **Baclofen** - titrate up slowly, I start with 2.5 mg TID for younger children, 5 mg TID for older children, 10 mg TID for adult-size, double the dose in a week

- **Other options**: Diazepam (Valium), Tizanidine (Zaniflex), Dantrolene

- Useful when spasticity causes discomfort and is widespread (quadriplegia)
- **Sedation** is the limiting side effect
Oral Medication: dystonia

- **Trihexyphenidyl (Artane)**- Titrate up slowly to 0.5-1 mg/kg/day divided TID. Watch for constipation.

- **Levodopa-carbidopa (Sinemet)**- titrate up slowly to 10 mg/kg/day divided TID. Can have stomach upset.

- **Baclofen** – mechanism in dystonia unclear

*Limited evidence for efficacy*
Botulinum Toxin

- Used to treat focal spasticity and dystonia
- Causes muscle weakness
- IM injections, lasts 3-6 months
- Useful when there are limited areas requiring treatment and functional goals. E.g.:
  - Calf muscles for flat-footed gait
  - Hip adductors for decreased scissoring, or ease of diapering/hygiene
  - Elbow flexors for better arm extension
Selective Dorsal Rhizotomy

• Dorsal spinal cord roots are selectively cut
• Interrupts the reflex arc, decreasing spasticity
• Typically used to treat spasticity in ambulatory patients with spastic diplegic cerebral palsy.
• Results in decrease tone, improvements in gait
• Limitations of the procedure:
  • Weak patients may lose function
  • Irreversible
Selective Dorsal Rhizotomy

The innervation pattern of each dorsal root is examined with EMG. A rootlet is sectioned.
Intrathecal Baclofen (ITB) Pump

• Delivery of baclofen into the spinal fluid space.
• Greater effect using very small doses (mcg), decreased sedating side effects
• Indicated for severe spasticity
• May be tried for dystonia
• Trial to predict effect
• ITB withdrawal is life-threatening, but high satisfaction rate (80%)
Tone Management: Approach

• Establish goals
• Identify tone abnormality, distinguish from contracture
• If few muscles groups involved, consider botulinum toxin
• If many muscle groups and discomfort or significant impairment from tone: try oral spasticity medications. If this fails, consider baclofen pump.
• Spastic diplegia: consider dorsal rhizotomy
Cases
Case 1

• 8 yo boy with mixed quadriplegic CP
  – Difficulty sleeping at night
  – Hard to get him into his wheel chair because of arching
  – Difficulty with dressing because of tightness
Case 1

• Severe increased tone (spasticity and dystonia) with significant QOL impact
• Oral baclofen tried, though too sleepy
• Successful ITB trial
• Baclofen pump placed with good effect
Case 2

- 8 yo girl with dyskinetic cerebral palsy
  - Non-ambulatory
  - Some tightness in the morning causing some discomfort
  - She has limited arm use due to abnormal movements
Case 2

• She came to me on oral baclofen. She was more irritable off, so it was continued
• Discussed possible use of artane
• Could also consider ITB therapy, however family feels she has good QOL and does not want to pursue this
Case 3

• 10 yo girl with left hemiplegic CP
  – Walks on her left toe
  – Occasional falls
  – Some pain at the end of the day
  – Does not tolerate AFO brace
Case 3

- Botox to left gastrocnemius and posterior tibialis
- Serial casting
Summary

• Identify the type of abnormal increased tone
• Establish goals
• Oral medication (baclofen) for discomfort relating to increased tone
• Botox for focal increased tone
• Rhizotomy for diplegia
• ITB therapy when oral baclofen fails
Additional Resources
