Cerebral Palsy:
Management of Complex Hypertonicity

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Objectives

- Learn about common types of tone abnormalities in cerebral palsy
- Considerations for medical management of hypertonicity, including spasticity and dystonia
- When to consider referring a patient for surgical interventions
- How to identify good candidates for selective dorsal rhizotomy, baclofen pump, or deep brain stimulation

Review: Cerebral Palsy Diagnosis

- Cerebral palsy is due to both genetic and acquired causes
- Accurate classification is important to direct early interventions and maximize functional outcomes
- Cerebral palsy is more than motor symptoms- is it important to recognize all symptoms that are impacting function and quality of life
- Early diagnosis can enable interventions during critical window of neuroplasticity in early life
- There are several tools that can be used for early diagnosis including neuroimaging, assessment of general movements, and the HINE

Tone Evaluation

Motor Examination

- Three parts to motor exam:
  1. Strength
  2. Bulk
  3. **Tone**

Definition of Tone

The resistance felt by the examiner during passive stretching of a joint when the muscles are at rest

Classification of Tone

1. Normotonic


2. Hypotonic – relaxed or "floppy" muscles


3. Hypertonic

Hypertonicity

1. Spasticity
2. Dystonia
3. Rigidity
4. Intrinsic Hypertonia (due to increased muscle connective tissue)

LOVE WILL.

Holland Bloorview Kids Rehabilitation Hospital

Bloorview RESEARCH INSTITUTE

HYPERTONIA ASSESSMENT TOOL (HAT) - SCORING CHART

Name:_____ **Chart/File #:**_____

Clinical Diagnosis:_____ **Date of Birth:** _____

Limb Assessed: **Gender:** ☐ Male ☐ Female

☐ Arm ☐ Left ☐ Right **HAT Assessor:**_____

☐ Leg ☐ Left ☐ Right **Date of Assessment:** _____

HYPERTONIA ASSESSMENT TOOL (HAT)

HAT ITEM	SCORING GUIDELINES (0=negative or 1=positive)	SCORE 0=negative 1=positive *(circle score)*	TYPE OF HYPERTONIA
1. Increased involuntary movements/postures of the designated limb with tactile stimulus of another body part	0= No involuntary movements or postures observed	0	DYSTONIA
	1= Involuntary movements or postures observed	1	
2. Increased involuntary movements/postures with purposeful movements of another body part	0= No involuntary movements or postures observed	0	DYSTONIA
	1= Involuntary movements or postures observed	1	
3. Velocity dependent resistance to stretch	0= No increased resistance noticed during fast stretch compared to slow stretch	0	SPASTICITY
	1= Increased resistance noticed during fast stretch compared to slow stretch	1	
4. Presence of a spastic catch	0= No spastic catch noted	0	SPASTICITY
	1= Spastic catch noted	1	
5. Equal resistance to passive stretch during bi-directional movement of a joint	0= Equal resistance not noted with bi-directional movement	0	RIGIDITY
	1= Equal resistance noted with bi-directional movement	1	
6. Increased tone with movement of another body part	0= No increased tone noted with purposeful movement	0	DYSTONIA
	1= Greater tone noted with purposeful movement	1	
7. Maintenance of limb position after passive movement	0= Limb returns (partially or fully) to original position	0	RIGIDITY
	1= Limb remains in final position of stretch	1	

SUMMARY SCORE – HAT DIAGNOSIS

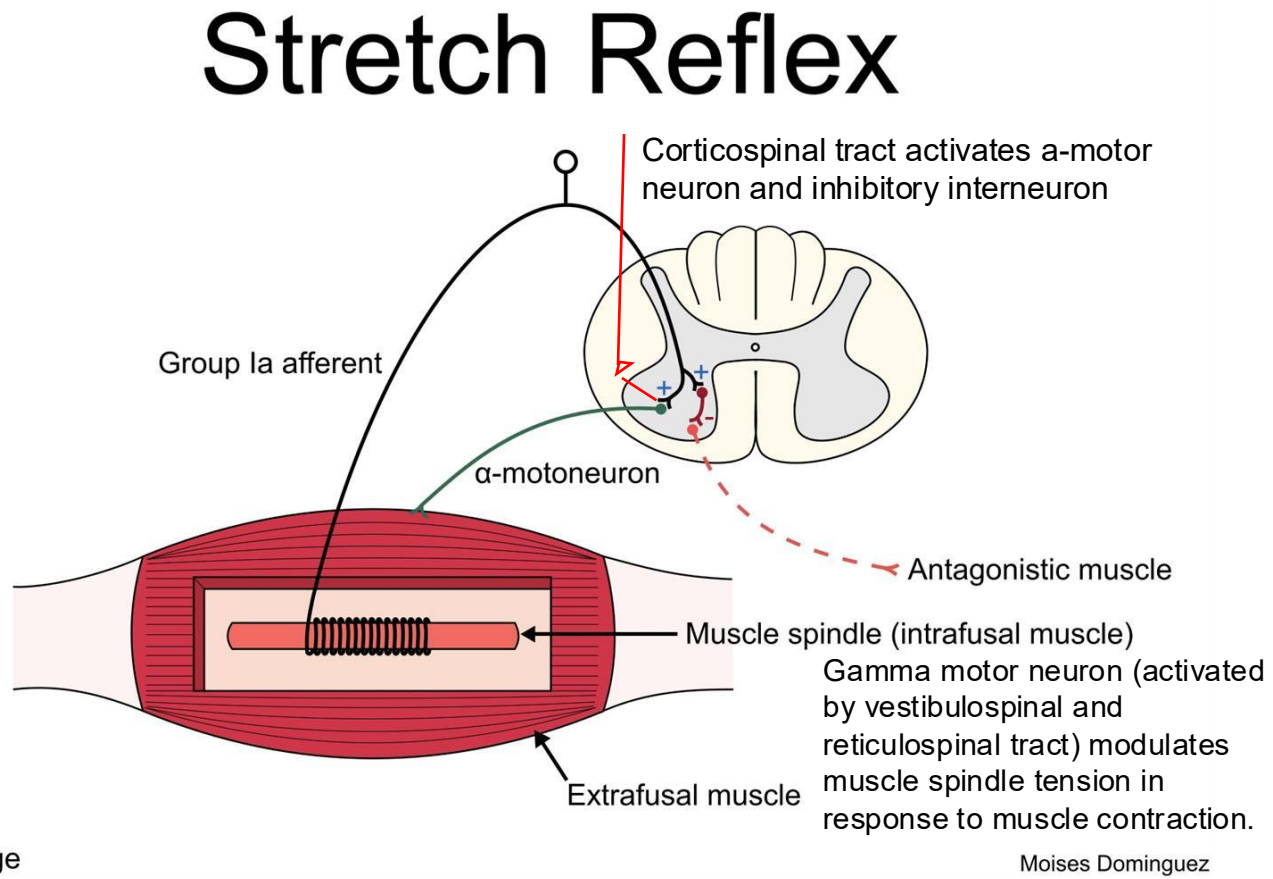
		Check box:
DYSTONIA →	Positive score (1) on at least one of the Items #1, 2, or 6	☐ Yes ☐ No
SPASTICITY →	Positive score (1) on either one or both of the Items #3 or 4	☐ Yes ☐ No
RIGIDITY →	Positive score (1) on either one or both of the Items #5 or 7	☐ Yes ☐ No
MIXED TONE →	Presence of 1 or more subgroups (e.g. dystonia, spasticity, rigidity)	☐ Yes ☐ No

HAT DIAGNOSIS: ***(Fill in all that apply)*** _____

Spasticity

- *"A disorder of the sensorimotor system characterized by a **velocity-dependent** increase in tonic **stretch reflexes** ('muscle tone') with exaggerated tendon jerks, resulting from hyperexcitability of the stretch reflex, as one component of the **upper motor neuron** syndrome."*

-JW Lance 1980



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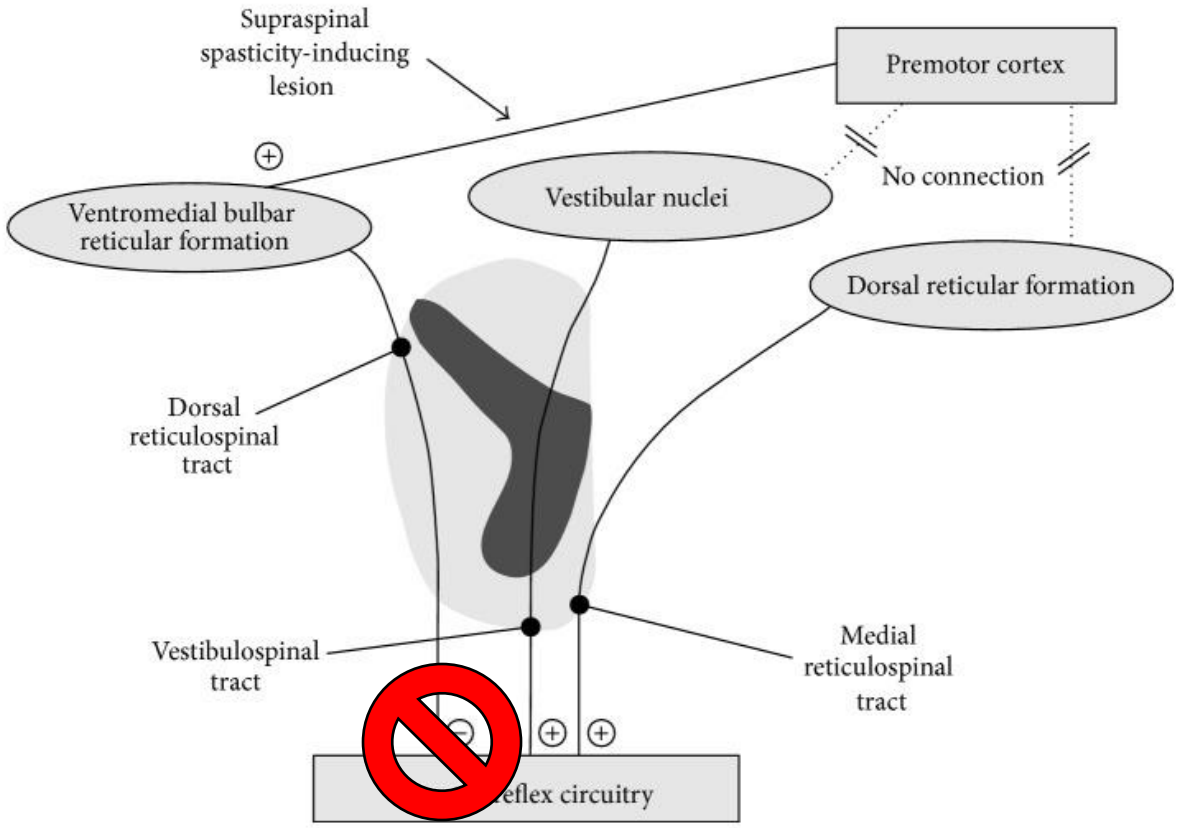

Spasticity- HAT

3. Velocity dependent resistance to stretch	0= No increased resistance noticed during fast stretch compared to slow stretch	0	SPASTICITY
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4. Presence of a spastic catch	0= No spastic catch noted	0	SPASTICITY
	1= Spastic catch noted	1	

Modified Ashworth Scale

Score	
0	No increase in muscle tone
1	Slight increase in muscle tone, manifested by a catch and release (tone normalization) or by minimal resistance at the end of the range of motion when the affected part(s) is moved in flexion or extension. Catch is a sudden slight increase in a muscle tone at any point during the range of motion (ROM) in the joint
1+	Slight increase in muscle tone, manifested by a catch (without release), followed by minimal resistance throughout the remainder (less than half) of the ROM
2	More marked increase in muscle tone through most of the ROM, but affect part(s) easily moved
3	Considerable increase in muscle tone passive, movement difficult
4	Affected part(s) rigid in flexion or extension

Rigidity

- Continuous involuntary sustained muscle contraction
- When an affected muscle is passively stretched, the degree of resistance remains constant regardless of the rate at which the muscle is stretched. This feature helps to distinguish rigidity from muscle spasticity.
- Seen as a feature of parkinsonian symptoms (decreased dopamine). Uncommon in children.

Rigidity- HAT

5. Equal resistance to passive stretch during bi-directional movement of a joint	0= Equal resistance not noted with bi-directional movement	0	RIGIDITY
	1= Equal resistance noted with bi-directional movement	1	
7. Maintenance of limb position after passive movement	0= Limb returns (partially or fully) to original position	0	RIGIDITY
	1= Limb remains in final position of stretch	1	

Dystonia

“Dystonia is a movement disorder characterized by sustained or intermittent abnormal movements, postures, or both. Dystonic movements and postures are typically patterned and repetitive and may be tremulous or jerky. They are often initiated or worsened by voluntary action, and frequently associated with overflow movements."

-Albanese, *Mov Disord*, 2025

Dystonia Characteristics

- Variable
- Often triggered by environmental or intrinsic stressors
- Worsens with attempts at voluntary movement
- Often presents with overflow activation of other muscle groups
- Improves during sleep

Dystonia- HAT

1. Increased involuntary movements/postures of the designated limb with tactile stimulus of another body part	0= No involuntary movements or postures observed	0	DYSTONIA
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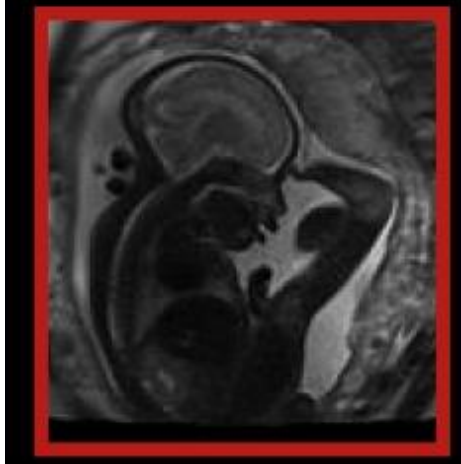
Dystonia Example

- 15-year-old boy presenting with slowly progressing twisting movements of his trunk, hands, and feet.

https://players.brightcove.net/6056665225001/default_default/index.html?videoId=6138658214001

Which is normal?

A



Fetus 35 week
Gestation
Absent corpus callosum
Normal subsequent devlopment

B

Healthy
6 month-old
infant

C

7 year old
DYT-1 mutation
Positive dystonia

Lin JP, Front Neurol, 2016

Dystonia Pathophysiology

- Thought to be due to imbalance between dopaminergic and cholinergic neurotransmitters in brain
- Dystonia isn't due to damage or dysfunction of any particular structure, but is rather a disorder of motor networks

Annals of NEUROLOGY
An Official Journal of the American Neurological Association and the Child Neurology Society
AMERICAN NEUROLOGICAL ASSOCIATION

Research Article | Full Access

Network Localization of Pediatric Lesion-Induced Dystonia

Rose Gelineau-Morel MD, Nomazulu Dlamini MD, PhD, Joel Bruss BA, Alexander L. Cohen MD, PhD, Amanda Robertson MSc, Dimitrios Alexopoulos MS,, Christopher D. Smyser MD, Aaron D. Boes MD, PhD

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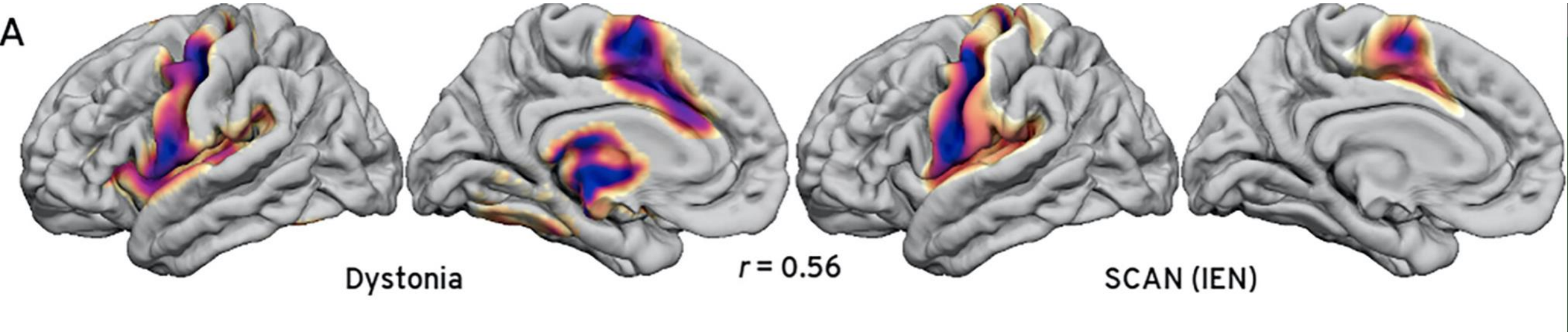

Hypertonicity Management

Medical Management

Medication Management

- Limited evidence to guide medication selection and dosing, especially in children
- Guidelines are largely based on expert opinion

Oral Medications for Movement Disorders

- For outpatient initiation: start low and titrate to the lowest effective dose for symptom control with tolerable side effects. To minimize side effects, start with divided dose given daily and titrate to 2-4 times daily [depending on half-life].
- Avoid abrupt discontinuation, unless otherwise noted. Most medications can be weaned every 3 to 7 days.
- Avoid all herbal supplements, especially St. Johns Wort, Ginko Biloba, Gotu Cola, Valerian Root, Kava Kava, SamE

Medications categorized by general indication in pediatric hypertonicity	
Dystonia, chorea, athetosis	**Spasticity**
Benzodiazepines	Baclofen
Carbamazepine/oxcarbazepine (data much stronger for carbamazepine, but oxcarbazepine has favorable side effect profile)	Benzodiazepines
Carbidopa/levodopa (dopa-responsive)	Cyclobenzaprine
Clonidine	Dantrolene
Levetiracetam	Gabapentin
Tetrabenazine	Metaxalone
Trihexyphenidyl	Pregabalin
Valbenazine	Tizanidine

Spasticity Management

Practice Parameter: Pharmacologic treatment of spasticity in children and adolescents with cerebral palsy (an evidence-based review)

M. R. Delgado, MD, FRCPC, FAAN, D. Hirtz, MD, FAAN, M. Aisen, MD, FAAN, S. Ashwal, MD, FAAN, D. L. Fehlings, MD, MSc, FRCPC, J. McLaughlin, MD, L. A. Morrison, MD, M. W. Shrader, MD, A. Tilton, MD, FAAN, and J. Vargus-Adams, MD, MS | AUTHORS INFO & AFFILIATIONS

January 26, 2010 issue • 74 (4) 336-343 • https://doi.org/10.1212/WNL.0b013e3181cbcd2f

- Consider if patient is using spasticity to help with function prior to treatment (e.g. in patient with weakness)
- Reasons to treat spasticity:
  - reducing pain and muscle spasms
  - facilitating brace use
  - improving posture
  - minimizing contractures and deformity
  - facilitating mobility and dexterity
  - improving patient ease of care as well as hygiene/self-care

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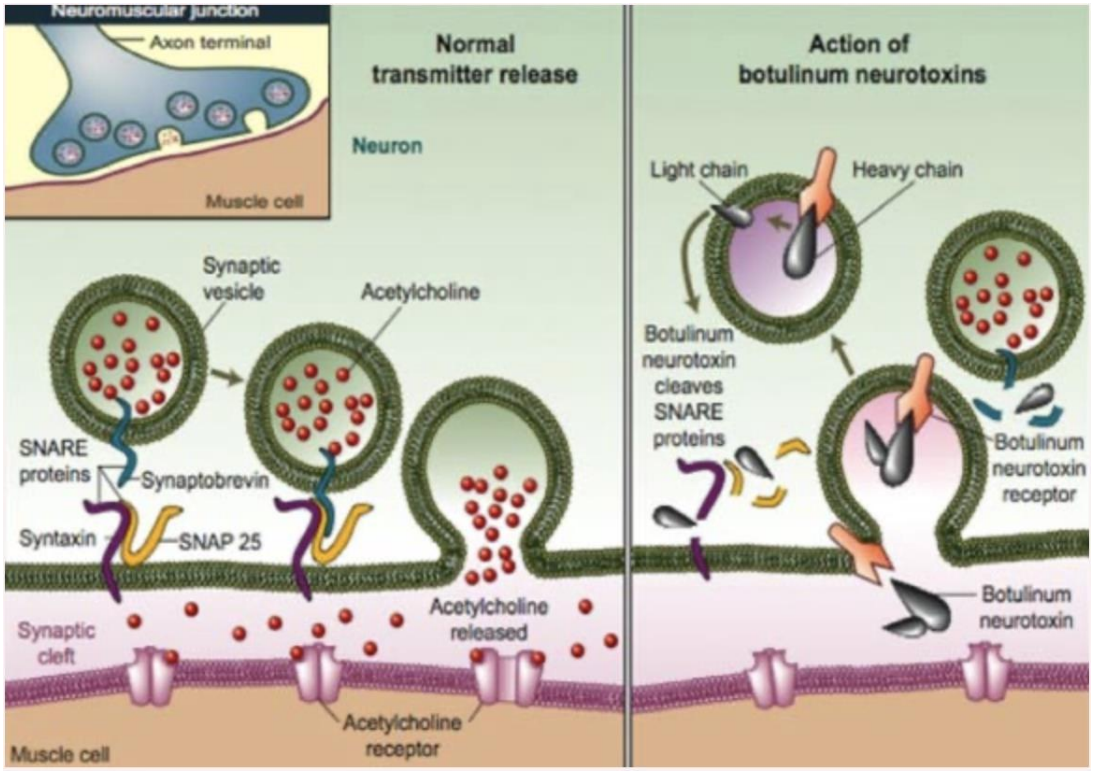
- Localized/segmental spasticity
  - Botulinum toxin type A is recommended (Level A)
  - Insufficient evidence to support phenol, alcohol, or botulinum toxin type B (Level U)

Botulinum Toxin



- Focal
- Reversible
- Minimal systemic effects
- May cause muscle atrophy if used long term

- BoNT-A --> binds SNAP 25
- BoNT-B --> binds synaptobrevin

Al-Ghamdi 2015

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- Generalized spasticity
  - Short term use of Diazepam (Level B)
  - Tizanidine can be considered (Level C)
  - Insufficient evidence for dantrolene, oral baclofen or intrathecal baclofen (Level U)

*In clinical practice, oral baclofen is frequently used first-line for spasticity treatment

Dystonia Management

"The major unmet need of children with dystonia, especially those with dystonia secondary to structural brain lesions or to metabolic disorders, is the availability of effective symptomatic treatment.…because dystonia seems to result from disordered function of complex neural circuits it is likely that dysfunction arising at any one (or several) node in those circuits may result in similar manifestations. Yet, ***effective treatment may differ across disorders depending on which node is primarily affected****."*

Mink, Mov Disord, 2013

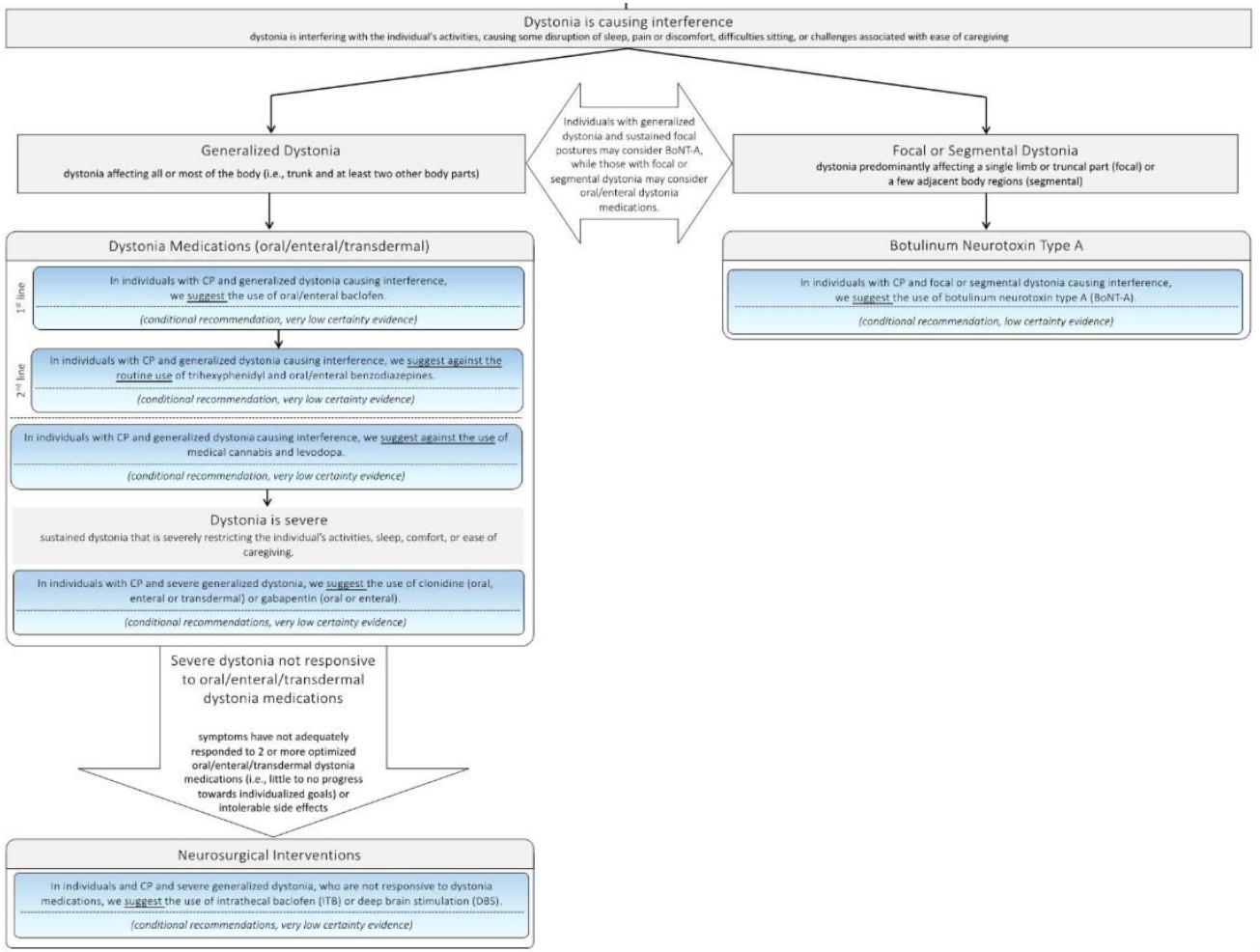
AACPDM Dystonia Care Pathway

- Optimize general health to treat dystonia triggers
- Assess impact of dystonia
  - Pain/discomfort
  - Activities of daily living
  - Ease of caregiving
  - Quality of life
- Determine individual intervention goals
- Introduce rehabilitation strategies
  - Physical/Occupational therapy
  - Bracing/Equipment

https://www.aacpdm.org/publications/care-pathways/dystonia-in-cerebral-palsy

AACPDM Dystonia Care Pathway

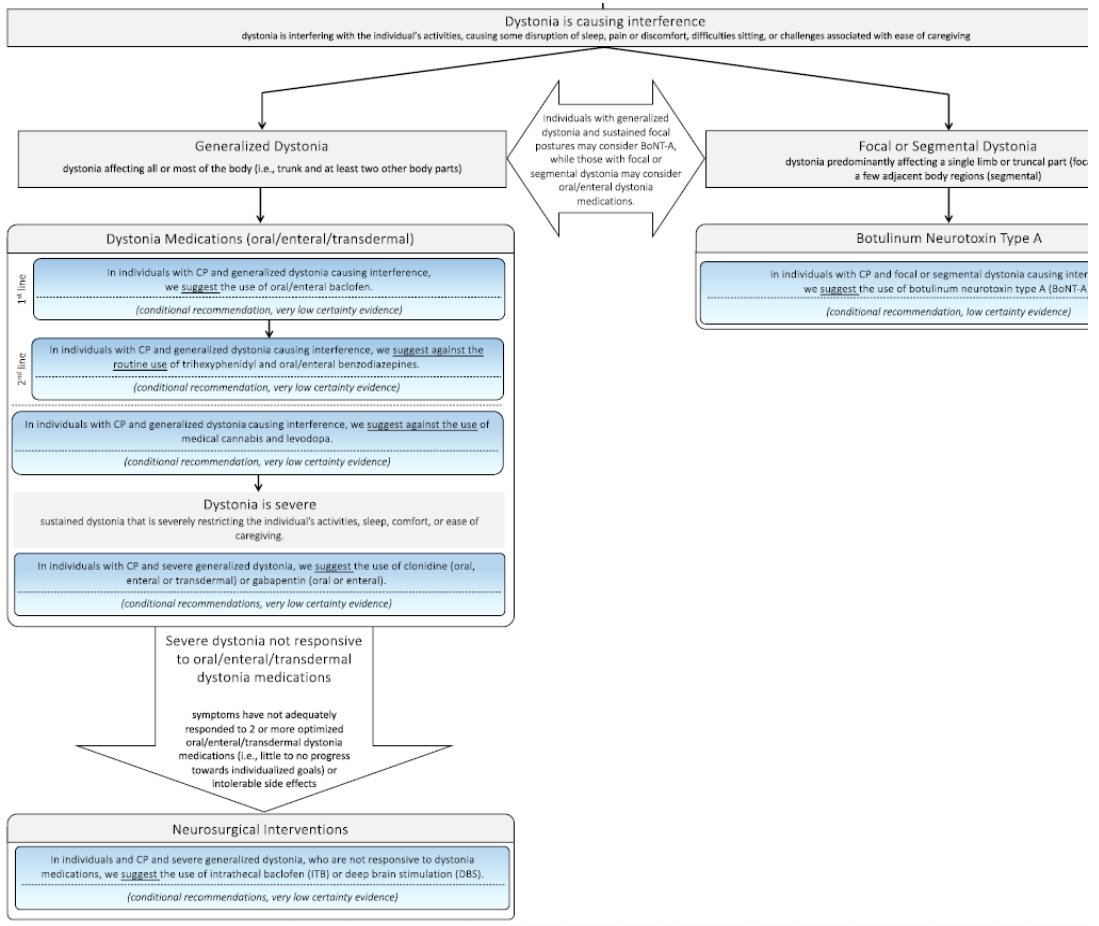
- Focal/Segmental Dystonia
  1. BoNT-A
- Generalized Dystonia
  1. Oral baclofen
  2. Trihexyphenidyl (caution for side effects)
  3. Oral benzodiazepines (caution for side effects)
  4. Levodopa
  5. Medical cannabis



AACPDM Dystonia Care Pathway

- Severe Dystonia
  1. Clonidine
  2. Gabapentin
- Not responding to 2 or more optimized medications
  1. Intrathecal baclofen pump
  2. Deep brain stimulation

https://www.aacpdm.org/publications/care-pathways/dystonia-in-cerebral-palsy



Additional Treatment Considerations

- Consider medical co-morbidities when selecting medication
  - Baclofen or benzodiazepines for comorbid spasticity
  - Clonidine to help with sleep
  - Gabapentin for painful dystonia
  - Trihexyphenidyl to help with sialorrhea
- When adding additional medications, consider trialing other mechanisms of action

Genetic dystonia

- Start with levodopa trial, titrating up to ~5mg/kg/day (can increase further if responsive)
- Levodopa is typically non-sedating
  - May need to have last dose of around 2PM to avoid sleep disturbance
- Trihexyphenidyl
  - Alone or in combination with levodopa
  - Consider if a lot of trouble with secretions
  - Titrate slowly to effect/side effects (doses up to 120mg/day have been reported)

Dystonia "spells"

- Treat the underlying cause!
- Consider clonidine, gabapentin, etc to target underlying dysautonomia that may trigger spells
- Can also add a dystonia rescue medication (generally benzos and muscle relaxants are best)
  - Clonazepam bridge x 3 days in illness
  - Intranasal midazolam or ODT clonazepam good for more acute rescue
  - Tizanidine can also be used as needed
- If no response to rescue plan/worsening symptoms, admit and follow status dystonicus guidelines

Status Dystonicus

Step 1: Admit to ICU/floor based on clinical picture and treat triggers and metabolic disturbances

- IV hydration
- Antipyretics
- Analgesia
- Promote comfort and sleep to break cycle
- Initiate dystonia rescue plan (if established)

Step 2: Optimize dystonia medications

- Focus on "heavy hitting" medications such as baclofen, benzodiazepine, muscle relaxants, gabapentin
- Also consider complementary medications with different MOA, such as Sinemet, trihexyphenidyl, and tetrabenazine
- Use higher than normal starting dosing and titrate quickly over days rather than weeks (anticipate concurrent side effects)
- Treat dysautonomia with clonidine, gabapentin, etc
  - Bromocriptine helpful for treating hyperthermia

Step 3: Parenteral/ Invasive treatments to promote sleep/comfort

- IV midazolam
- IV Precedex (alpha 2 agonist similar to clonidine)
- IV phenobarbital
- IV propofol (limit to <2 days if possible)
- Paralytic agents (non-depolarizing such as pancuronium)
- Consider invasive therapy for refractory cases (baclofen pump, DBS, pallidotomy, non-selective rhizotomy, etc)

- Recognition of status dystonicus: can't tolerate lying, disturbed sleep
- Progresses to metabolic disturbances: fever, dehydration, abnormal electrolytes, CK >1000, myoglobinuria

Adapted from Allen et al, Dev Med and Child Neuro, 2013

Hypertonicity Management

Surgical Management

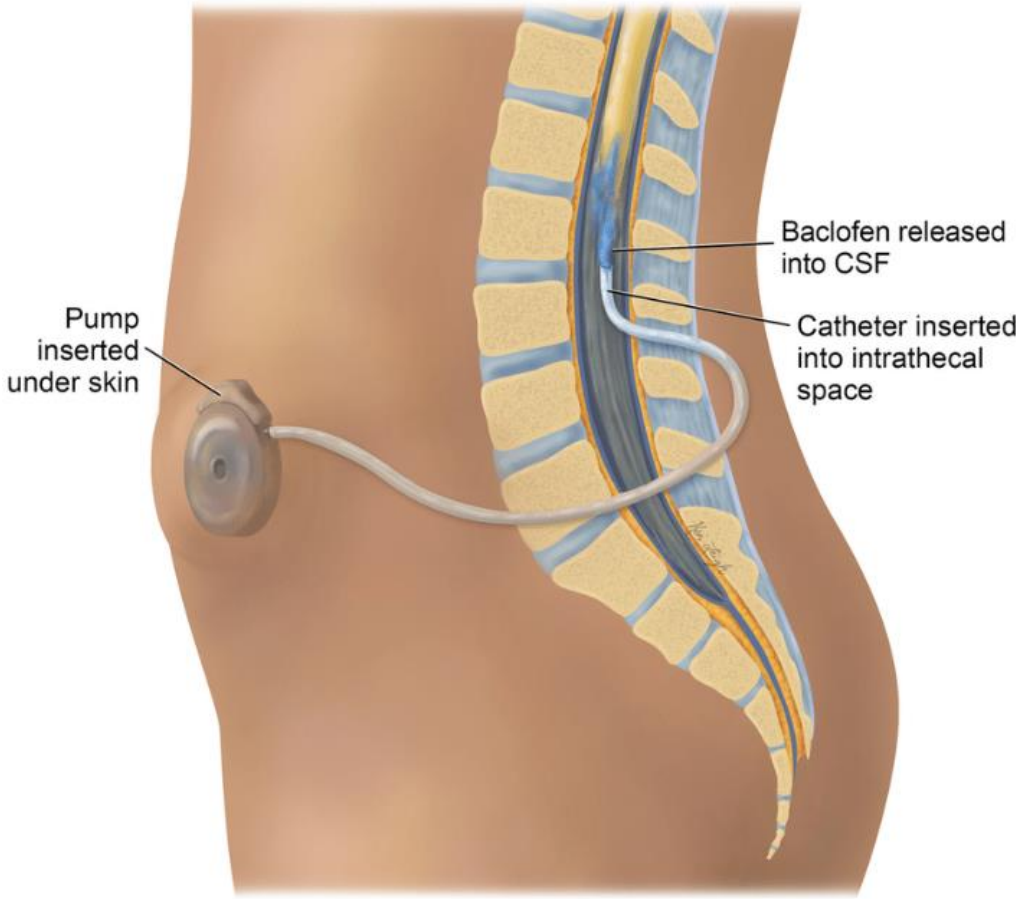
Surgical Management of Hypertonicity

- Patients who have failed 2 or more medications for treatment

OR

- Diagnosis with known good response to surgical option
  - E.g., Deep brain stimulation for DYT-TOR1A

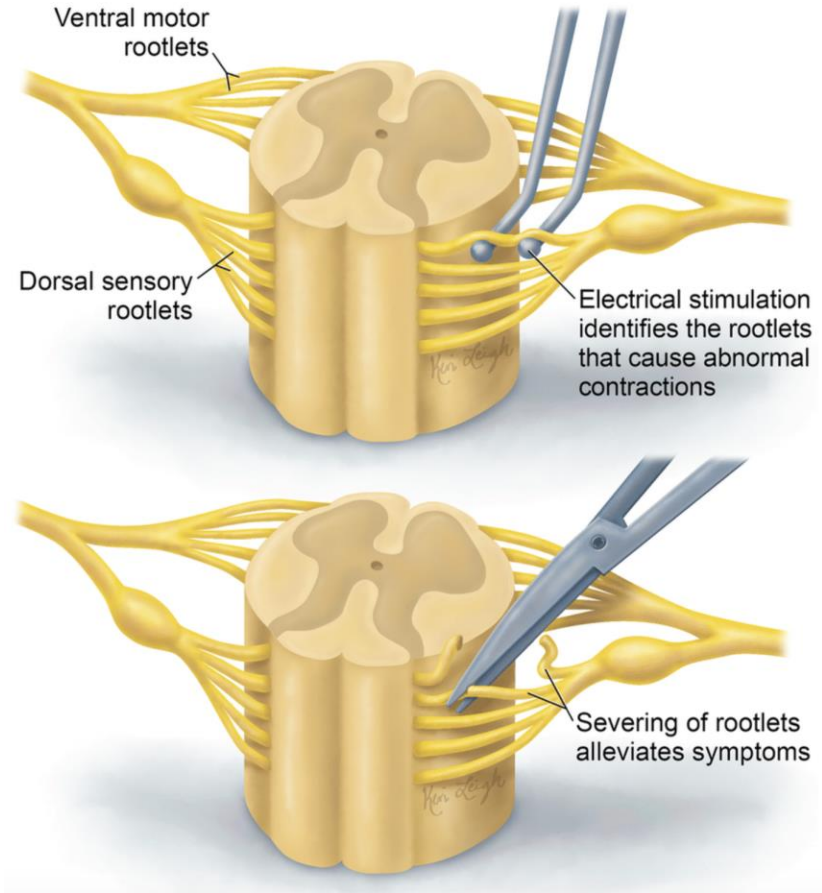
Intrathecal Baclofen Pump



Kudva et al, Neurosurgical Review, 2021.

- Delivers baclofen to spinal column
- Gives much smaller dose than oral ($1/100^{th}$ dose)
- Lower extremities > Upper extremities
- Reversible
- Risk of baclofen withdrawal/ pump failure
- Consider in patients with mixed spasticity and dystonia

Selective Dorsal Rhizotomy

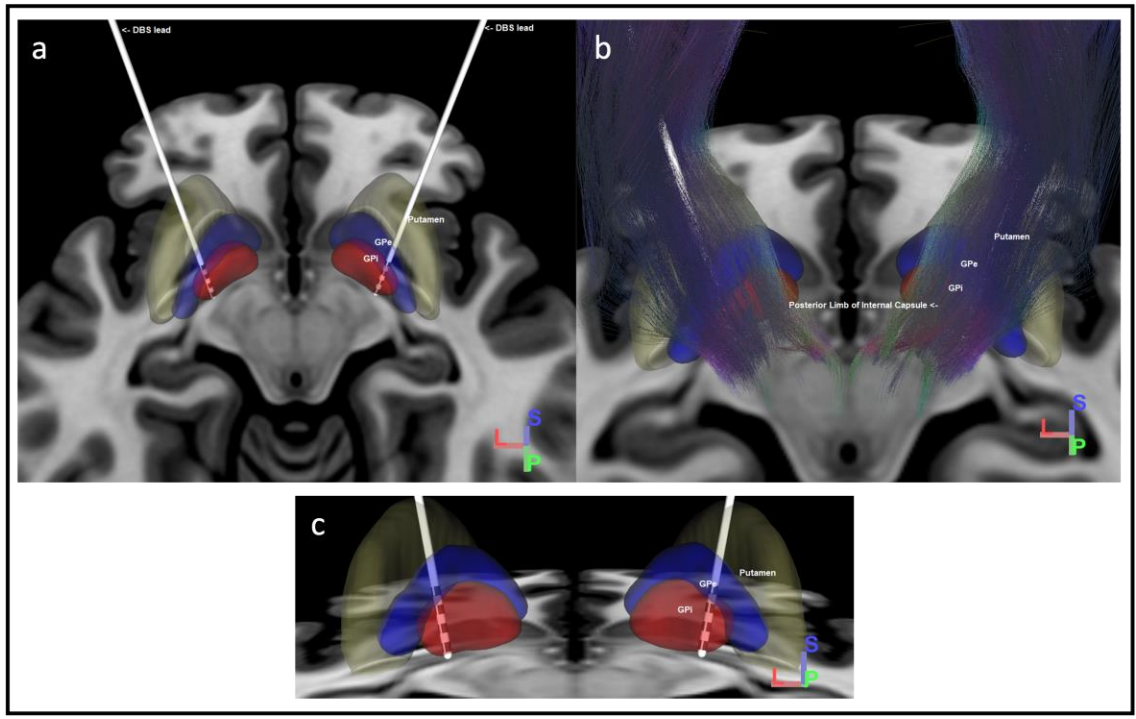


Kudva et al, Neurosurgical Review, 2021.

- Non-reversible
- No risk of withdrawal or equipment infection
- Only use for patients with minimal/no dystonia
  - Can unmask dystonia with relief of spasticity and lead to worse functional outcome than prior to surgery

Deep Brain Stimulation

- Consider for patients with monogenic dystonia not responsive to multiple medications, or for certain patients with acquired dystonia
- Can consider as first line in some monogenic dystonias (DYT-TOR1A)



- Patients >7 years old
- Shorter duration of dystonia predictive of better response
- Measure response over months, not weeks

Other surgical options

- Dorsal ventral rhizotomy/palliative rhizotomy
  - Consider for patients with severe dystonia who failed baclofen pump
  - Irreversible
  - Causes weakness
- Pallidotomy
  - Irreversible
  - Only treats dystonia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