A ‘Care Pathway’ for Dystonia in Cerebral Palsy

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Objectives

- Understand the definition of dystonia and how it helps in the classification of the CP sub-type (NIH Taskforce on Childhood Motor Disorders)
- Gain knowledge in diagnosing dystonia using the HAT (Hypertonia Assessment Tool)
- Introduction to the AACPDM ‘Dystonia in CP” Care Pathway
NIH Taskforce on Childhood Motor Disorders

Classification and Definition of Disorders Causing Hypertonia in Childhood
Terence D. Sanger, Mauricio R. Delgado, Deborah Gaepler-Spira, Mark Hallett and Jonathan W. Mink
*Pediatrics* 2003:111:e89-e97

Definition and Classification of Negative Motor Signs in Childhood
Terence D. Sanger, Daofen Chen, Mauricio R. Delgado, Deborah Gaepler-Spira, Mark Hallett, Jonathan W. Mink and the Taskforce on Childhood Motor Disorders
*Pediatrics* 2006;118:2150-2167
DOI: 10.1542/peds.2005-3016

Definition and Classification of Hyperkinetic Movement Disorders in Childhood
Terence Sanger, Daofen Chen, Darcy Fehlings, Mark Hallett, Anthony Lang; Jon Mink and Taskforce on Childhood Motor Disorders

Movement Disorders
*Movement Disorders*
Vol. 25, No. 11, 2010, pp. 1538–1549
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- Hypertonia: defined as ‘abnormally increased resistance to passive stretch’
- Three types of neurologically mediated hypertonia: spasticity, dystonia, and rigidity
- Dystonia is ‘a movement disorder in which involuntary sustained or intermittent muscle contractions cause twisting and repetitive movements, abnormal postures or both’
Spasticity is hypertonia in which ‘resistance to externally imposed movement increases with increasing speed of stretch and/or resistance to externally imposed movement rises rapidly above a threshold speed or angle’

Rigidity is ‘velocity-independent bidirectional resistance which may involve simultaneous co-contraction of agonists and antagonists’

Mixed tone: occurs when more than one sub-type of hypertonia co-exist

Dystonia is ‘a movement disorder in which involuntary sustained or intermittent muscle contractions cause twisting and repetitive movements, abnormal postures or both’
**Chorea: Definition**

“Is an ongoing random-appearing sequence of one or more discrete involuntary movements or movement fragments”

**Athetosis: Definition**

“Is a slow continuous, involuntary writhing movement that prevents the maintenance of a stable posture”
Hallmark Features of Dystonia in CP

- Dystonia is both an involuntary movement disorder and a cause of hypertonia
- Tone is variable
- Stiff movement
- Sustained postures (often twisting)
- Involuntary movements/postures/tone are triggered by voluntary movement, excitement, tactile stimuli

Key Features of Cerebral Palsy

- Group of motor disorders (abnormal tone, posture, control of movement)
- Onset before during or after birth (usually before 3 years of age)
- Secondary to a brain injury or anomaly
- Non-progressive and permanent
“Definition” of Cerebral Palsy
Rosenbaum et al DMCN 2007, S109:8-14

• Group of disorders of the development of movement and posture causing an activity limitation
• Attributed to non-progressive disturbances that have occurred in the developing brain
• Are often accompanied by disturbances of sensation, cognition, communication, perception, and/or behaviour and/or seizure disorder

Interaction between Components of the ICF - WHO 2001

Health Condition
(disorder/disease)

Impairment
(function/structure)

Activities
(task/action)

Participation
(involvement in life situation)

ENVIRONMENTAL FACTORS

PERSONAL FACTORS
Sub-types of CP

Cerebral Palsy

- Spastic
  - Bilateral
  - Unilateral
  - Diplegic
  - Quadriplegic

- Dyskinetic
  - Hypokinetic
  - Hyperkinetic

- Ataxic
  - Hemiplegic
  - Mixed CP: combination of subtypes

Dystonic
Choreoathetosis
• Numerous scales exist to “quantify” the severity of hypertonia (e.g. Tardieu, MAS, ASA, BAD)
• A standardized clinical tool to differentiate the different forms of hypertonia does not exist
• Current standard is the neurological examination (lacks standardization and is experience dependent)
• Potential Uses of the HAT:
  – 1) Clinical: to guide treatment (e.g. medication type and dose may depend on the type of hypertonia)
  – 2) Research: to “describe” subjects

3 Clinical Exam Manoeuvres to Diagnose Dystonia:
- tactile stimulation of a body part to trigger postures
- voluntary movement of a distant body part to trigger postures
- increased tone with voluntary movement of a distant body part
Why Treat Dystonia in CP?

- Improve motor function
- More commonly we aim to relieve pain associated with dystonia or decrease tone to facilitate care-giving
Characteristics of Pain in Children and Youth With Cerebral Palsy
Melanie Pennet, Wen Yan Xue, Navneet Bainspal, Lauren Switzer and Darcy Fehlings
DOI: 10.1542/peds.2013-0224

Holland Bloorview
Kids Rehabilitation Hospital

Treatments for Dystonia

- Rehabilitation Treatments
- Oral Medications
- Botulinum Toxin
- Intrathecal Baclofen
- Deep Brain Stimulation
Dystonia Care Pathway AACPDM Site


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Flow Diagram for an Evidence-Informed Care Pathway for Dystonia in Cerebral Palsy

**Assessment**
- Determine if dystonia is present (e.g., HTT, neuro exam) and if generalized or focal
- Assess severity of dystonia (history, neuro exam)
- Determine if dystonia is consistent with CP diagnosis (risk factors, brain imaging) or if additional work up is required (e.g., MRI of brain)
- Assess impact of dystonia on function, quality of life

**Periodic re-evaluation of dystonia**

**Determine intervention goals for:**
- Dystonia causing pain
- Dystonia interfering with function

**Manage secondary health issues (nutrition, GI tract, sleep)**
Slide Organization: Oral Medications

Drug/Main Category of Usage

- Mechanism of Action
- Dosage
- Side Effects
- Efficacy/Effectiveness
- Evidence
Oral Pharmacotherapy for Dystonia

- anticholinergic
- dopaminergic

Oral Pharmacotherapy: Baclofen

- Binds GABA$_B$ receptors of spinal interneurons presynaptically
- Dosage: work up to 2mg/kg po divided tid to qid
- Side Effects: drowsiness (can be minimized by increasing the dosage slowly)
- Efficacy: moderate reduction in dystonia initially but tolerance can develop
- Evidence: U
Oral Pharmacotherapy for Dystonia:
Trihexyphenidyl (Artane)

- **Mechanism of Action:** anticholinergic (suppresses an excess of cholinergic activity present in dystonia)
- **Dosage:**
  - Starting Dosage: 0.5 mg bid (<15 kg) and 1.0 mg bid (>15 kg) [available in a 2 mg tablet] – can work up to 0.5 to 0.75 mg/kg/day
  - Usual dosage range: 1 to 10 mg bid to qid
- **Side effects:** dry mouth, nausea, blurred vision, urinary retention, drug rash, chorea
- **Efficacy:** mixed results, can help with drooling!
- **Evidence:** U

Oral Pharmacotherapy for Dystonia:
Carbidopa/L-Dopa

- **Mechanism of Action:** Dopaminergic
- **Dosage:** Available as Sinemet e.g.100/25 (one tablet contains 100 mg levodopa and 25 mg carbidopa); 1-2 tablets bid to tid
- **Side effect:** nausea (most common), other side effects include a decrease in blood pressure, depression, tremor, chorea, psychosis
- **Efficacy:** Treatment of Choice in Dopamine Responsive Dystonia (can be misdiagnosed with CP)
Specific Indications

- Gabapentin for pain relief with dystonia
- Benzodiazepines/Clonidine for dystonia storms

Oral Medications for Dystonia: General Comments

- Potential role in generalized dystonia (versus focal dystonia)
- Significant variability amongst health care providers in the use of oral meds
- Minimal information on the long-term cognitive effects of the oral medications
- Care Pathway: reserve for children with generalized dystonia or where dystonia is causing pain/sleep disturbance
Conclusion

- Dystonia is a frequent cause of hypertonia and involuntary movements in children with CP
- Dystonia is characterized by “stiff postures”
- Dystonia is a frequent cause of pain in children with CP
- Dystonia in CP can be diagnosed using the HAT
- Majority of the AACPDM Care Pathway for Dystonia in CP is based on clinical expert opinion – more evidence is required