Spasticity and Gait Disorders in Adults and Children with Hydrocephalus

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Gait

- One’s manner of walking
- **Walking** — a controlled fall
- **Locomotion** — rhythmical limb and trunk movements that propel a person
- **Balance** — postural responses that permit a person to attain and maintain appropriate posture for locomotion and protect against falls when the body is not in equilibrium because of voluntary actions or perturbations arising from outside the body (anticipatory or reactive responses)
What’s Necessary for Gait and Balance?

- **Balance clues**
  - Vestibular input, vision, proprioception

- **Physical capability**
  - Motor system (cortex, cerebral white matter, spinal cord, peripheral nerves, muscles)
  - Functioning bones and joints, minimal pain, cardiopulmonary system

- **Coordination, control, and motivation**
  - Cerebellum, basal ganglia, frontal lobes, goals
Etiologies of Nonneurologic vs. Neurologic Gait Impairment

Nonneurologic
- Arthritis, cardiac disease, chronic lung disease, and peripheral vascular disease
- Low vision a common contributor

Neurologic
- Unsteady, ataxic, frontal, parkinsonian, neuropathic, hemiparetic, and spastic

Patients may have both
- i.e., multifactorial gait impairment
Common Neurologic Causes

Brain
- Stroke, chronic subcortical ischemia, movement disorders (e.g., parkinsonism), hydrocephalus, degenerative dementias

Spinal cord
- Myelopathy (e.g., cervical or lumbar stenosis)

Nerve
- Neuropathy (e.g., diabetes)

Muscle
- Myopathy, myasthenia gravis
History

- Difficulty getting in and out of low seats, the toilet, or the car
- Difficulty initiating gait (hesitation)
- Shuffling, scuffing, tripping, limping, falling
- Touching walls or furniture, or holding on for balance; fearfulnessness
- Unsteadiness on turns
- Avoidance or difficulty with stairs, curbs, ramps, uneven surfaces
General Neurologic Exam

- **Strength/weakness patterns**
  - Hemiplegia, monoplegia, general weakness
  - Rigidity, tremor, paratonia, flaccidity

- **Sensory patterns**
  - Distal sensory loss, hemi-sensory

- **Reflexes**
  - Asymmetry or hyperreflexia, clonus, spasticity

- **Cerebellar**
  - Ataxia, dysarthria
Examination of Gait

To evaluate gait, physician should walk with patient
Good to get into a hallway
Physician should watch patient walk
Factors to watch
  - Getting in and out of chair, maintenance of stance and balance, initiation of gait, pace, cadence, stride length, stability on turns, walking on toes or heels, tandem gait
Details of Gait Cycle

Figure 1. Timing of events during just over one complete gait cycle starting with right initial contact.

Australian Family Physician 2007; 36:398–405
Lower (simpler)

Middle (intermediate)

Higher (complex and integrative)
Lower-Level Gait Disorders

- Disturbances of **force production**
  - Myopathies, motor neuropathies, motor neuron disease
  - Foot drop, waddling gait

- Disturbances of **sensation**
  - Proprioception, vision, and vestibular
  - Sensory ataxia

- Easily elicited sensory and motor findings

Nutt JG, Lang AE. Balance and Gait Disorders Syllabus. 2011 American Academy of Neurology
Middle-Level Gait Disorders

- Impaired modulation of force generated by the lower-level motor system
  - Spasticity from disruption of corticospinal tracts
  - Ataxia from disturbances of the cerebellum and its connections
  - Hyperkinetic gait associated with chorea and dystonia
  - Hypokinetic gait associated with parkinsonism
    - Parkinsonian gait also has higher-level features

Nutt JG, Lang AE. Balance and Gait Disorders Syllabus. 2011 American Academy of Neurology
Higher-Level Gait Disorders

- Difficulties integrating sensory information about position of the body in the environment and in the gravitational field, and selecting and executing appropriate motor plans to accomplish person’s intentions.

- Postural and locomotor responses are inappropriate or absent.

Nutt JG, Lang AE. Balance and Gait Disorders Syllabus. 2011 American Academy of Neurology
5 Features that Suggest a Higher-Level Gait Disorder

- Freezing of gait
- Absent or counterproductive postural responses
- Inappropriate anticipatory postural responses seen in transitions from sitting to standing
- Variability in gait patterns related to environmental and emotional cues
- Absence of neurologic signs that explain the gait pattern

J Neural Transm 2007; 114:1253–8
Higher-Level Gait Disorders

- Patients with higher-level gait patterns generally do not have findings on the neurologic exam that explain the gait pattern
  - No primary motor or sensory deficits

- Makes them difficult to diagnose

- Frontal cortex, subcortical connections to basal ganglia and brainstem, and the basal ganglia
  - Hydrocephalus, atypical parkinsonism, PSP, subcortical ischemia

Nutt JG, Lang AE. Balance and Gait Disorders Syllabus. 2011 American Academy of Neurology
Reasons To Use An Assistive Device

- Redistribute and unload a weight-bearing lower limb
- Improve balance/prevent falls
- Reduce lower-limb pain
- Provide sensory feedback
A THREE-POINT GAIT

- Tripod position
- Step 1: Both crutches and affected leg advance
- Step 2: Unaffected leg advances

B TWO-POINT GAIT

- Tripod position
- Step 1: Left crutch and right foot advance
- Step 2: Right crutch and left foot advance

C FOUR-POINT GAIT

- Tripod position
- Step 1: Right crutch advances
- Step 2: Left foot advances
- Step 3: Left crutch advances
- Step 4: Right foot advances

medical-dictionary.thefreedictionary.com
ankle-foot orthosis

knee-ankle-foot orthosis
Therapeutic Exercise

- Flexibility
- Coordination
- Strength
- Endurance
- Speed
- Conditioning
Spasticity Overview
Spasticity is a motor disorder 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.

— Lance, 1980
## Upper Motor Neuron Syndrome

### Positive Symptoms
- Spasticity
  - Hyper-reflexia
  - Clonus
  - Clasp knife
- Flexor/extensor spasm
- Dystonia
- Rigidity

### Negative Symptoms
- Decreased dexterity
- Weakness
- Paralysis
- Fatigability
- Slowness of movement
Etiologies

- Stroke
- Traumatic brain injury
- Multiple sclerosis
- Spinal cord injury
- Cerebral palsy
- Anoxia
- Neurodegenerative disease
Pathophysiology of Impairment after a Central Nervous System Lesion

Immediate consequences
- Paralysis
  - Immobilization in shortened position

Delayed consequences
- Damage to higher center
  - Spasticity
  - Rearrangement of spinal activity
  - Muscle overactivity

Adult Spasticity Overview

Gracies et al, 1997
Impact of Spastic Disorders

- Mobility
- Pain
- Sleeping patterns
- Affect and mood
- Self-care
- Disfigurement
- Self-esteem

- Sexual function
- Fatigue
- Contracture
- Increased risk of falls
- Pressure sores
- Poor orthotic fit
- Caregiver burden
Possible Advantages of Spasticity

- Maintains muscle tone
- Helps support circulatory function
- May prevent formation of deep vein thrombosis (DVT)
- May assist in activities of daily living, transfers, gait
<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 flexion or extension</td>
</tr>
<tr>
<td>2</td>
<td>More marked increase in tone, but limb 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>

Source: Ashworth, 1964
Adult Spasticity Overview

Common Clinical Patterns: Upper Limbs

- The Adducted/Internally Rotated Shoulder
- The Flexed Wrist
- The Pronated Forearm
- The Clenched Fist
- The Flexed Elbow
- The Thumb-in-Palm Deformity
Common Clinical Patterns: Lower Limbs

- Equinovarus
- Striatal Toe
- Extended Knee
- Flexed Knee
- Adducted Thighs
Rationale for Treatment

- If spasticity interferes with:
  - Functioning
  - Positioning
  - Comfort
  - Care

- If spasticity is not useful (e.g., during transfers)
- If treatment is expected to provide meaningful improvement
## Adult Spasticity Overview

### Treatment Goals

<table>
<thead>
<tr>
<th>Improved</th>
<th>Decreased</th>
</tr>
</thead>
<tbody>
<tr>
<td>ROM</td>
<td>Energy expenditure</td>
</tr>
<tr>
<td>Mobility</td>
<td>Spasm frequency</td>
</tr>
<tr>
<td>Gait</td>
<td>Pain</td>
</tr>
<tr>
<td>Orthotic fit</td>
<td>Caregiver burden</td>
</tr>
<tr>
<td>Positioning</td>
<td></td>
</tr>
<tr>
<td>Ease of hygiene</td>
<td></td>
</tr>
<tr>
<td>Cosmesis</td>
<td></td>
</tr>
</tbody>
</table>
The Spasticity Management Team

- Patient
- Family/caregiver
- Physician champion
  - Physiatrist
  - Orthopedic surgeon
  - Neurosurgeon
  - Neurologist
- Rehab Team
  - PT, OT, TR, SLP
  - Social worker
  - Psychologist
- Gait lab
- Wheelchair/seating vendor
- Orthotist
- Pedorthotist
- Podiatrist
- Coordinator/CM
Considerations in Treatment Decisions

- Severity
- Degree of underlying selective control
- Distribution
  - Diffuse vs. focal
- Locus of CNS injury
- Availability of care & support
  - Technical expertise
  - Reimbursement issues
- Chronicity
  - Acute vs. chronic
  - Wait 3–6 months before considering chemodenervation
- Comorbidities
  - General poor health
  - Cognitive or psychiatric impairment
- Potential for complications
Considerations in Treatment Decisions

- Contracture
  - Patient evaluation for passive stretching, tone reduction techniques, dynamic splinting
  - Temporary anesthetic nerve block

- Diagnostic tests
  - Must do EMG before neurolytic procedure
Chemoneurolysis with Phenol

Materials

- 5% aqueous solution
- Nerve stimulator for percutaneous stimulation
- Teflon-coated cannulated nerve-block needle
Target Tissue

- Major peripheral nerve
  - Limited number appropriate
  - Motor nerve or nerve with limited sensory distribution
  - Musculocutaneous – elbow flexion
  - Obturator – thigh adduction
- Motor point
  - Intramuscular nerve branch to muscle fascicles
Mechanism of Action

- Immediate local anesthetic effect
- Indiscriminate axonal loss lesion
- Long duration of effect

Efficacy

- 3–6-month duration of effect, sometimes longer
- Depends on dose used
- Number of muscles/motor points
- Extent of reinnervation from collateral sprouting
Phenol

Benefits
- Cheap
- Little local tissue irritation
- Reasonably long duration of action

Contraindications
- Poor general health
- Active infection
- Severe contractures
- Anticoagulation
OPERATION: IRAQI FREEDOM
Botulinum 'is WMD'
State Department spokesman: Lethal bio-toxin 'kills people, it kills people in large quantities'

The vial of botulinum bacteria discovered in Iraq by U.S. arms inspectors – which experts call the most poisonous substance known to man – is "a weapon of mass destruction," the State Department's top spokesman announced yesterday.

"Botulinum kills people, it kills people in large quantities. Botulinum is a weapon of mass destruction, yes," said State spokesman Richard Boucher," according to an Agence France-Presse report. "Anything that destroys on a massive scale is a weapon of mass destruction."
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FDA Approves Botox to Treat Frown Lines

FDA today announced the approval of Botulinum Toxin Type A (Botox Cosmetic) to temporarily improve the appearance of moderate to severe frown lines between the eyebrows (glabellar lines), a medical condition that is not serious. The product's manufacturer, Allergan, Inc., Irvine, California, is now allowed to market Botulinum Toxin Type A for this new indication.
FDA-approved Medical Indications for Botulinum Toxin Therapy

- Cervical dystonia
- Strabismus
- Blepharospasm
- Severe primary axillary hyperhydrosis
- Upper limb spasticity
- Migraine
Regulated Exocytosis
Multiple Neurotransmitters / Neuropeptides Released From Vesicle

Adapted from Trends in Cell Biology, July 1997.
Advantages of Botulinum Toxin

- Can target small muscles
- Titratable
- Relatively short duration of action
- No risk of adverse sensory nerve involvement
Sample Botulinum Toxin Type A (Allergan, Inc.) Doses in Adults
Superficial Flexor Muscles of Left Forearm

**Flexor Carpi Ulnaris**
- Brashear: 50 U
- Richardson: 30–75 U
- Simpson: 10–40 U
- Dunne: 25–60 U
- Grazko: 5–40 U
- Pierson: 40–130 U

**Flexor Carpi Radialis**
- Brashear: 50 U
- Richardson: 50–75 U
- Simpson: 15–60 U
- Dunne: 20–75 U
- Grazko: 5–40 U
- Pierson: 40–210 U

**Flexor Digitorum Superficialis**
- Brashear: 50 U
- Richardson: 75 U
- Dunne: 20–150 U
- Grazko: 5–20 U
- Pierson: 40–85 U

**Flexor Pollicis Longus**
- Brashear: +/–20 U
- Dunne: 10–20 U
- Pierson: 40 U
BTX-A: Clinical Effects in Spasticity

- Injected directly into overactive muscles
- Focal, temporary chemodenervation
- Onset usually within 24–72 hr; maximum effect at ~2 weeks*
- Clinical benefit usually >12 weeks; may be extended with adjunctive therapy
- Can be used in conjunction with other therapies (e.g., systemic medications, intrathecal baclofen pump, phenol/alcohol)

*Brin, 1997
Before Botulinum Injection
Most Frequently Asked Questions
Why Does Botulinum Toxin Kill You?

- Presynaptic receptor blockade irreversible
- Paralysis of respiratory muscles
Why Does Botulinum Toxin Wear Off?

- Collateral sprouting of uninvolved nerve terminals reinnervates the muscle.

- Process of collateral sprouting occurs over ~3 months.
Will Any Be Left Over for My Wrinkles?
ITB™ Therapy
(Intrathecal Baclofen Therapy)
What is ITB Therapy?

- Delivers a liquid form of baclofen (Lioresal Intrathecal) directly into the intrathecal space, where fluid flows around the spinal cord
- Relieves spasticity with a small amount of drug delivered via a programmable pump to where it is most effective in the spinal fluid
- FDA approved
  - 1992 – Spinal-origin spasticity
  - 1996 – Cerebral-origin spasticity
SynchroMed Infusion System Components

- **Pump**
  - Infuses drug at programmed rate

- **Catheter**
  - Delivers drug to intrathecal space of spinal cord

- **Programmer**
  - Allows for precise dosing
  - Allows for adjustable dosing

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**SynchroMed® II**

**N’Vision Programmer**
Intrathecal Baclofen
Indications

- Generalized spasticity
- Spasms/pain
- Oral medication failure
- Comfort
- Functional goals
- Reliable patient/caregiver
Intrathecal Baclofen Advantages

- Titratable
- Effective
  - 2-point Ashworth reduction for spinal origin
  - 1-point Ashworth reduction for cerebral origin
- Reversible
- Compatible
Intrathecal Baclofen
Disadvantages

- Up to 15% complication rate – infection, failure, catheter problems
- Requires periodic refills (every 2–6 months, depending on concentration used)
- Requires battery replacement every 5–7 years
- Caution to avoid bolus (overdose) or abrupt withdrawal
Steps of ITB Therapy

- Step 1: Screening test
- Step 2: Surgical procedure
- Step 3: Therapy maintenance
Step 1: Screening Test

- The screening test allows team to evaluate potential efficacy
- A positive response is required before pump implant
  - 86% cerebral origin (screening test)
  - 97% spinal origin (screening test)
- Screening procedure will last 6–8 hr
- Standard amount of drug is injected into intrathecal space by lumbar puncture
- Drug effect is temporary
Step 1: Screening Test

- Pre-injection PT evaluation
- Injection of medication, 50 μg
- Medical/nursing monitoring
- Serial PT evaluations after injection
  - Immediately
  - Every 1–2 hr
- Discharge home when back-to-baseline tone
Step 2: Surgical Procedure

- Anesthesia required
- Pump is surgically placed, usually just under the skin of the abdomen
  - Neurosurgeon, physiatrist
- Catheter is threaded beneath skin into intrathecal space
- Short acute hospital stay may be required
- Inpatient rehabilitation
Step 3: Therapy Maintenance

- Active patient involvement and cooperation
- Regularly scheduled visits for dose titration
- Outpatient PT/OT
- Pump refill
  - 30–45-min appointment
  - A needle is put through skin and into pump
  - Old medicine is removed and new medicine is put in
  - Pump may be reprogrammed
Potential Risks of ITB Therapy

- Most common side effects include loose muscles, drowsiness, nausea/vomiting headache, dizziness
- Overdose, although rare, could lead to respiratory depression, confusion, loss of consciousness, seizures, reversible coma; in extreme cases, may be life-threatening
- No specific antidote – physostigmine
- Complications, when they occur, are usually surgically related
- Abrupt discontinuation (withdrawal) can result in itching, labile blood pressure, high fever, altered mental status, spasticity, muscle rigidity; in rare cases, it has been fatal
Summary

- Phenol
  - Cheap, limited sites but strategic, effective, lack of titratability not a big problem, botox sparing

- Botulinum toxin
  - Expensive, “fine instrument,” safe, effective

- Intrathecal baclofen
  - More invasive, generalized effect, but titratable
Summary

- Interventional procedures are not stand-alone
- Must define goals of treatment
- All 3 interventions may be applicable to one patient
- Coordinated team necessary for optimal outcomes
THANK YOU