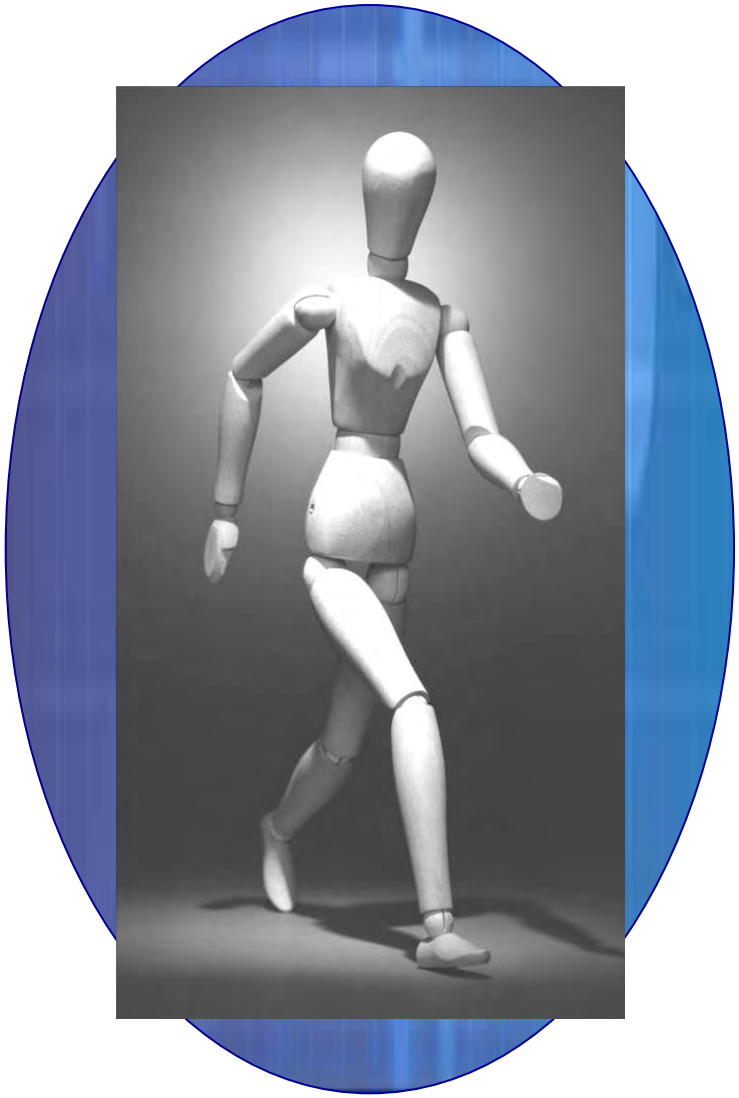

Spasticity and Gait Disorders in Adults and Children with Hydrocephalus

Scott E. Brown, MD

Chairman
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LIFEBRIDGE HEALTH℠
The Sandra and Malcolm Berman Brain & Spine Institute

SINAI HOSPITAL
a LifeBridge Health center
Rehabilitation Center

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?

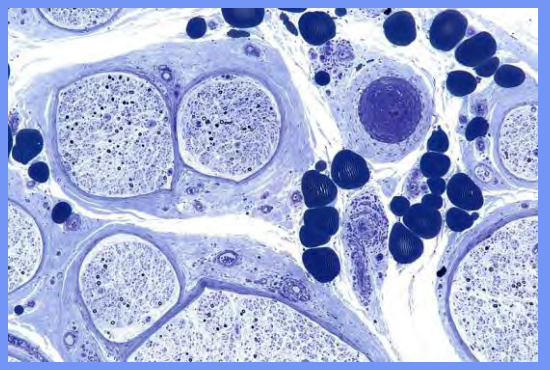
- Balance clues
  - Vestibular input, vision, proprioception
- Physical capability
  - Motor system (cortex, cerebral white matter, spinal cord, peripheral nerves, muscles)
  - Functioning bones and joints, minimal pain, cardiopulmonary system
- Coordination, control, and motivation
  - Cerebellum, basal 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 <u>both</u>
  - 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

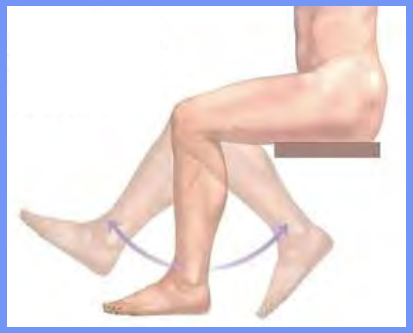


History

- 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; fearfulness
- 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

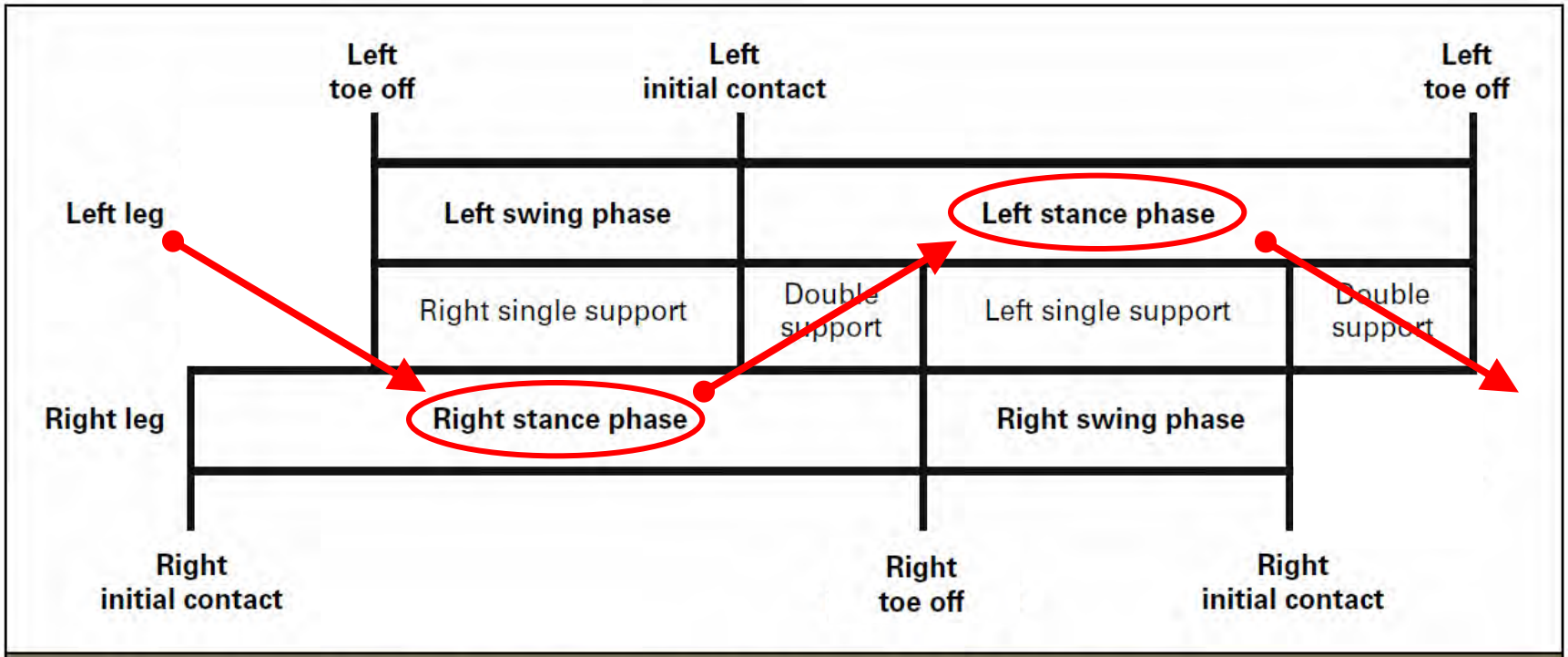


Figure 1. Timing of events during just over one complete gait cycle starting with right initial contact

Australian Family Physician 2007; 36:398–405

Gait Patterns Based on Neurologic Function

- Lower (simpler)
- Middle (intermediate)
- Higher (complex and integrative)

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

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

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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

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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

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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

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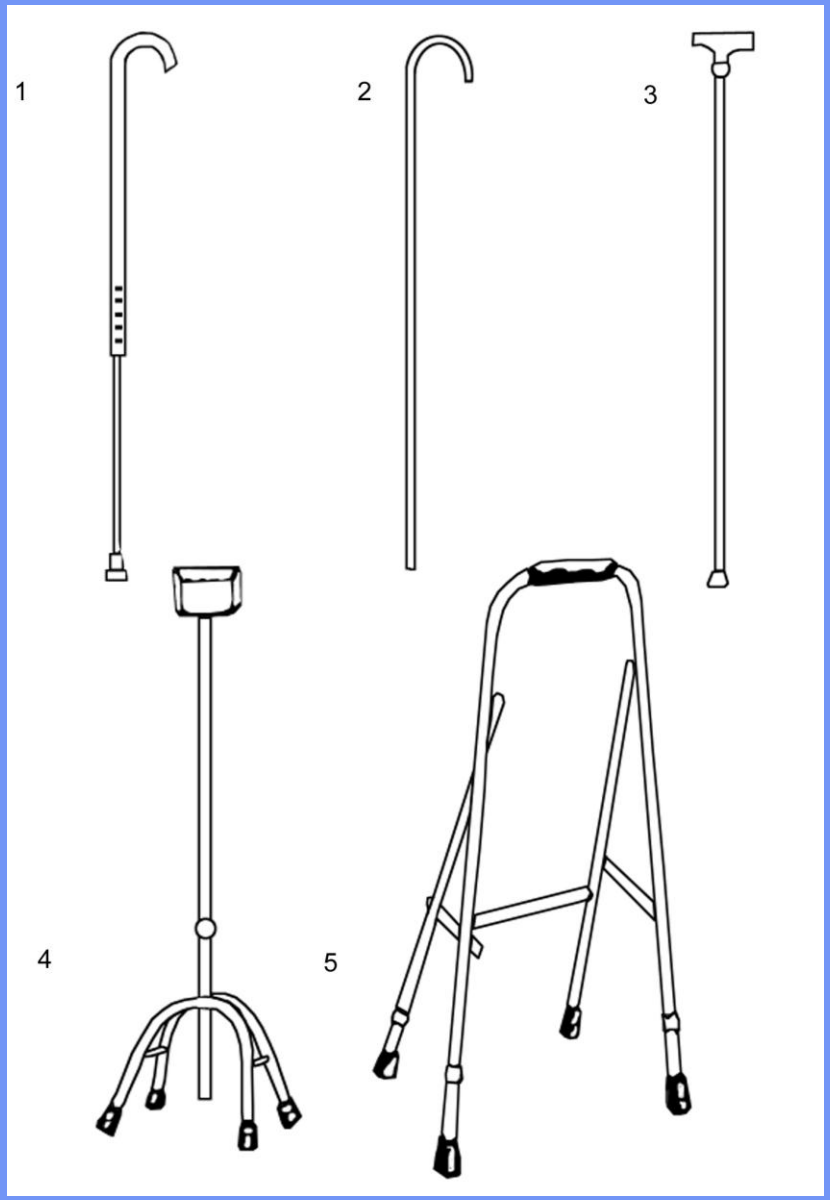
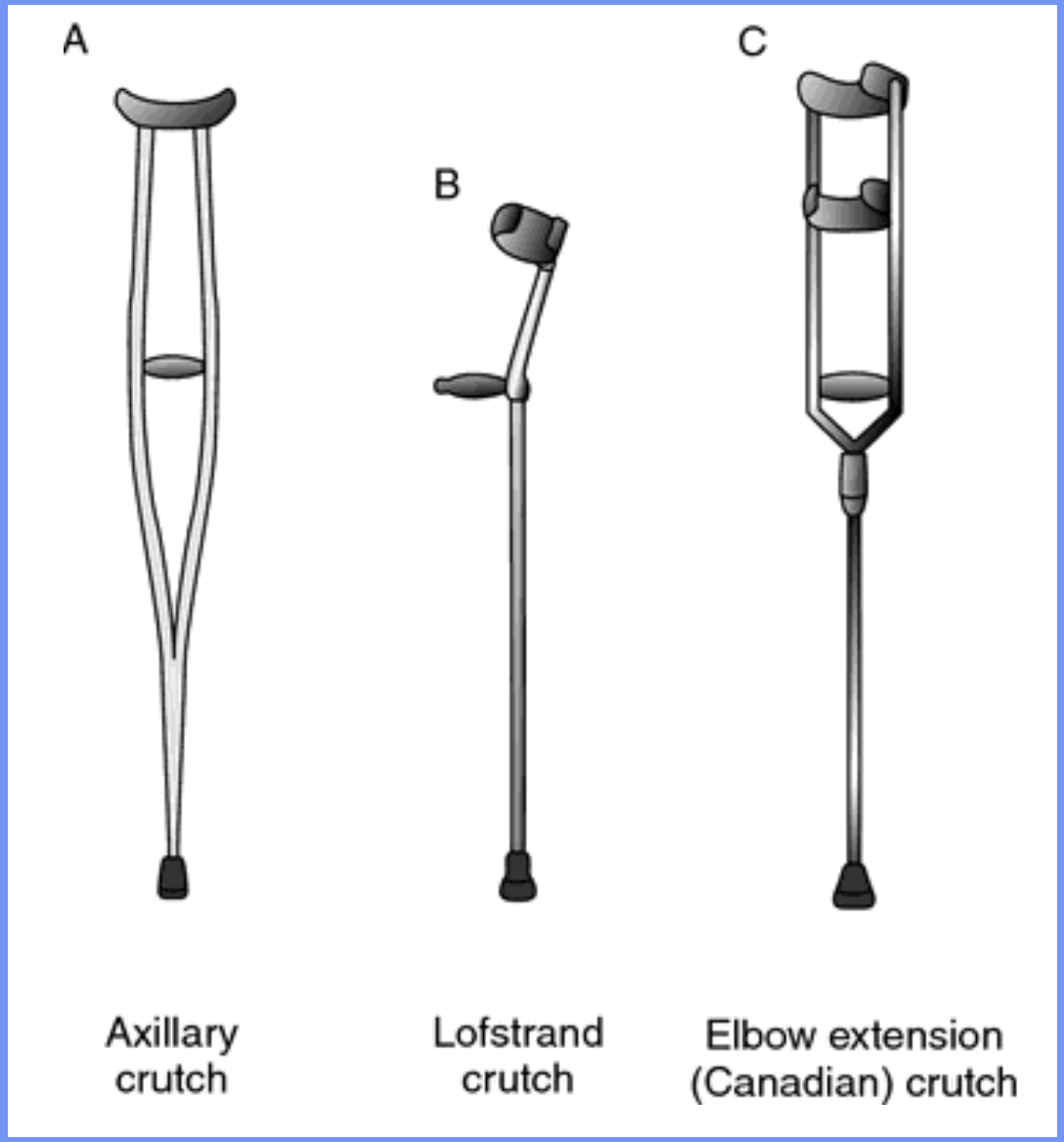


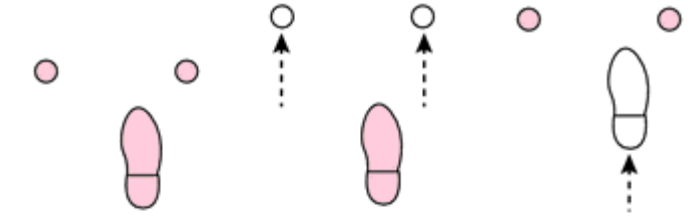
emedicine.medscape.com



medical-dictionary.thefreedictionary.com

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

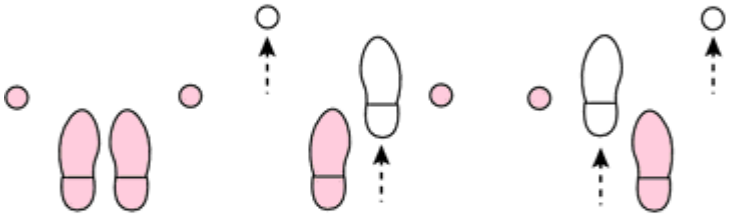


Tripod position

Step 1
Both crutches and affected leg advance

Step 2
Unaffected leg advances

A THREE-POINT GAIT

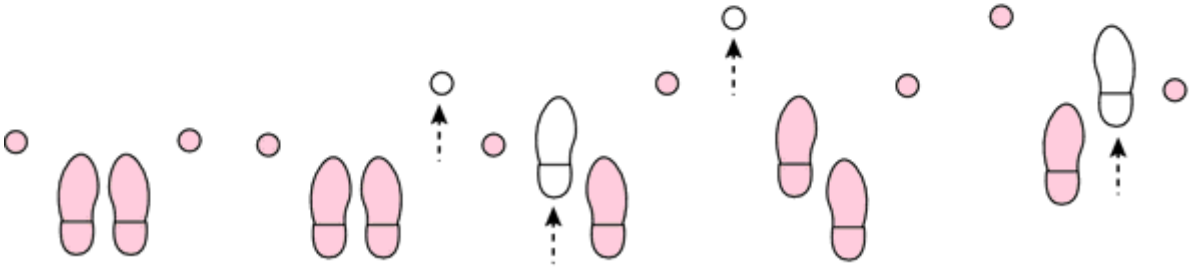


Tripod position

Step 1
Left crutch and right foot advance

Step 2
Right crutch and left foot advance

B TWO-POINT GAIT



Tripod position

Step 1
Right crutch advances

Step 2
Left foot advances

Step 3
Left crutch advances

Step 4
Right foot advances

C FOUR-POINT GAIT

ankle-foot orthosis

knee-ankle-foot orthosis

Therapeutic Exercise

- Flexibility
- Coordination
- Strength
- Endurance
- Speed
- Conditioning

Spasticity Overview

Definition of Spasticity

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

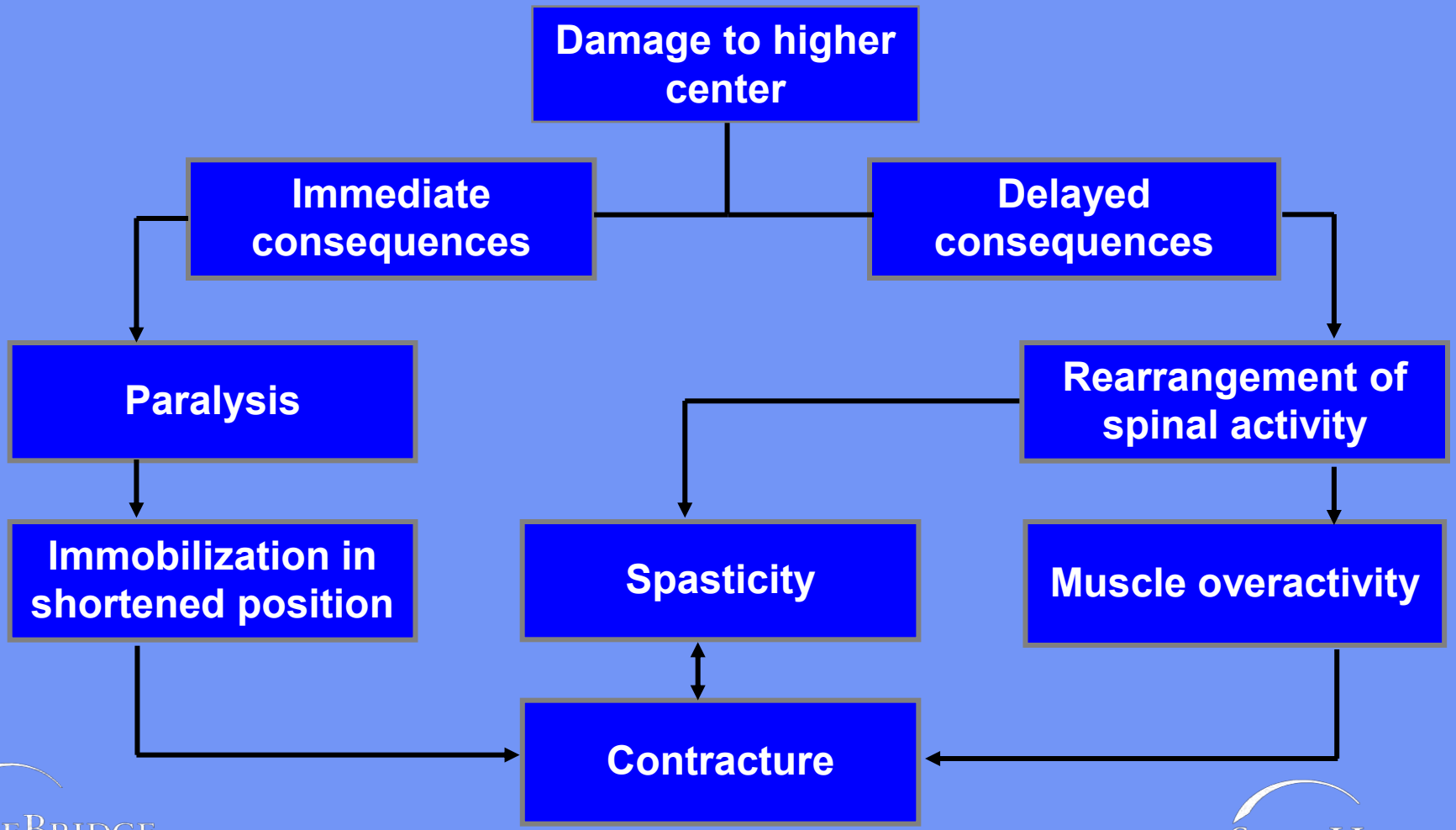
Negative Symptoms

- 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



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

Ashworth Scale

Grade	Description
0	No increase in tone
1	Slight increase in tone giving a “catch” when the limb is moved in flexion or extension
2	More marked increase in tone, but limb easily flexed
3	Considerable increase in tone, passive movement difficult
4	Limb rigid in flexion or extension

Source: Ashworth, 1964

Common Clinical Patterns: Upper Limbs

The Adducted/Internally Rotated Shoulder

The Flexed Wrist

The Pronated Forearm

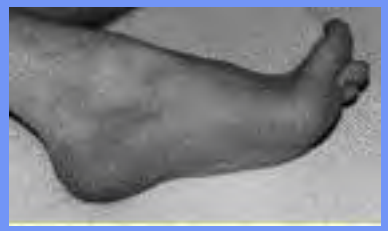
The Clenched Fist

The Flexed Elbow

The Thumb-in-Palm Deformity

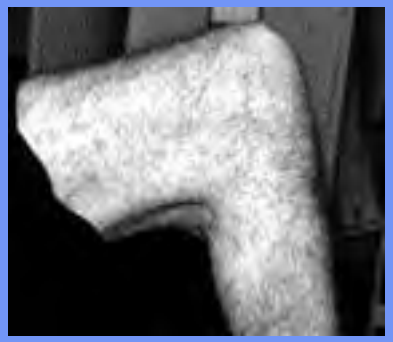
Common Clinical Patterns: Lower Limbs

Equinovarus

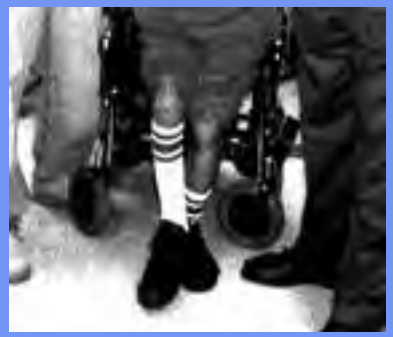


Striatal Toe

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

Treatment Goals

Improved	Decreased
ROM	Energy expenditure
Mobility	Spasm frequency
Gait	Pain
Orthotic fit	Caregiver burden
Positioning	
Ease of hygiene	
Cosmesis	

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

WorldNetDaily™
Saturday, October 4, 2003

OPERATION: IRAQI FREEDOM

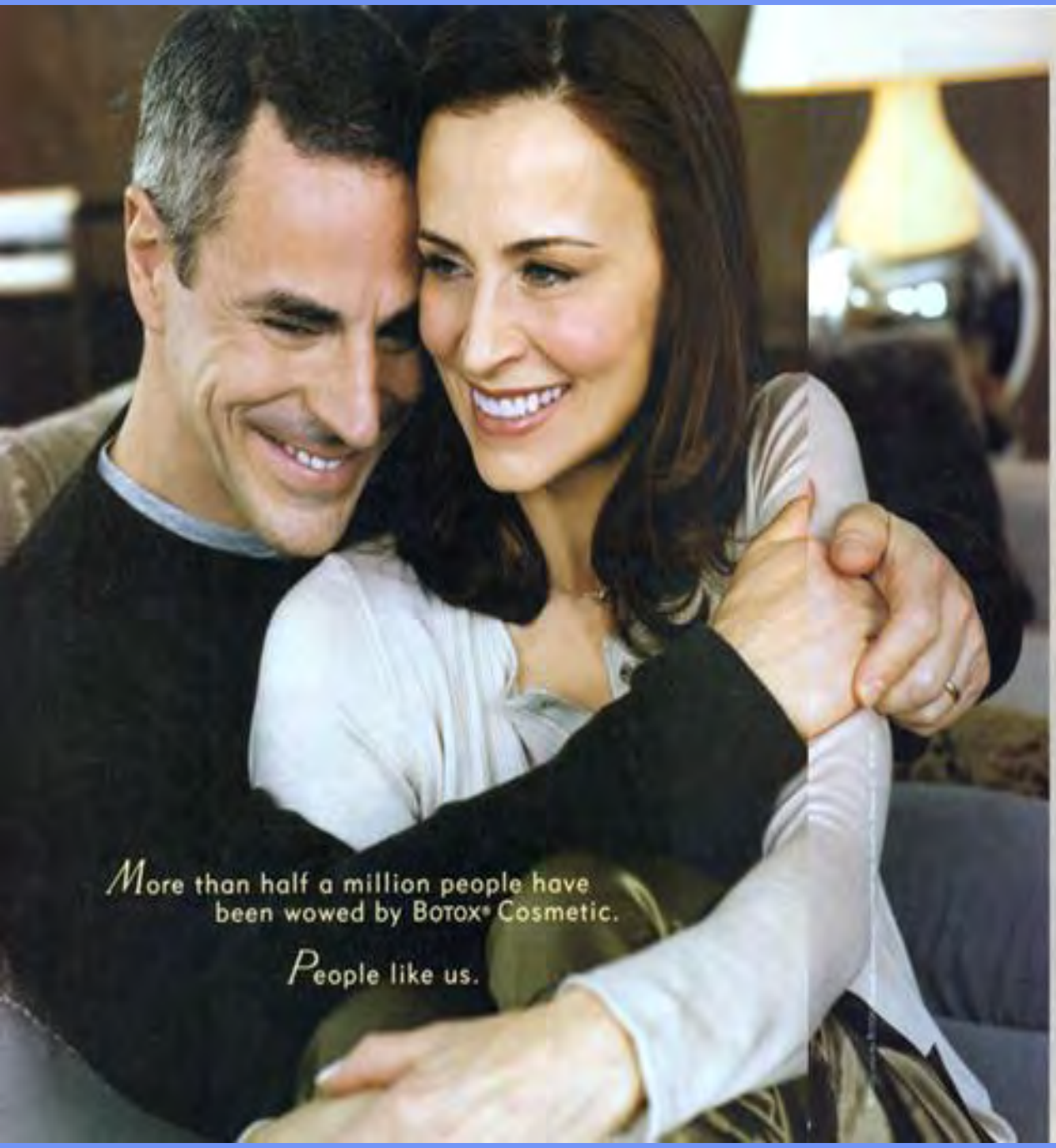
Botulinum 'is WMD'

State Department spokesman: Lethal bio-toxin 'kills people, it kills people in large quantities'

Posted: October 4, 2003
1:00 a.m. Eastern

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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U.S. Food and Drug Administration

FDA Talk Paper

FDA Talk Papers are prepared by the Press Office to guide FDA personnel in responding with consistency and accuracy to questions from the public on subjects of current interest. Talk Papers are subject to change as more information becomes available.

T02-20

April 15, 2002

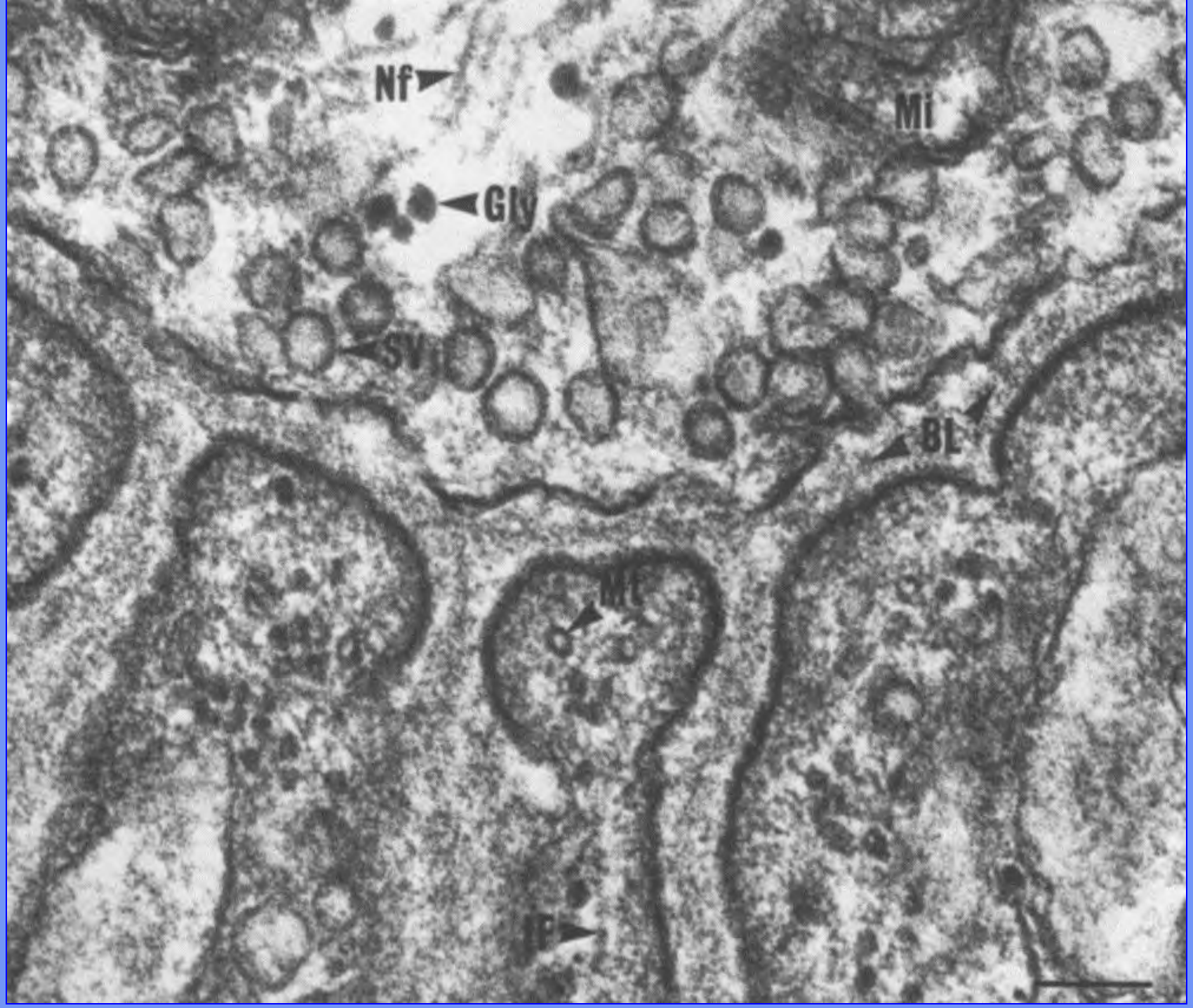
Media Inquiries: 301-827-6242
Consumer Inquiries: 888-INFO-FDA

FDA APPROVES BOTOX TO TREAT FROWN LINES

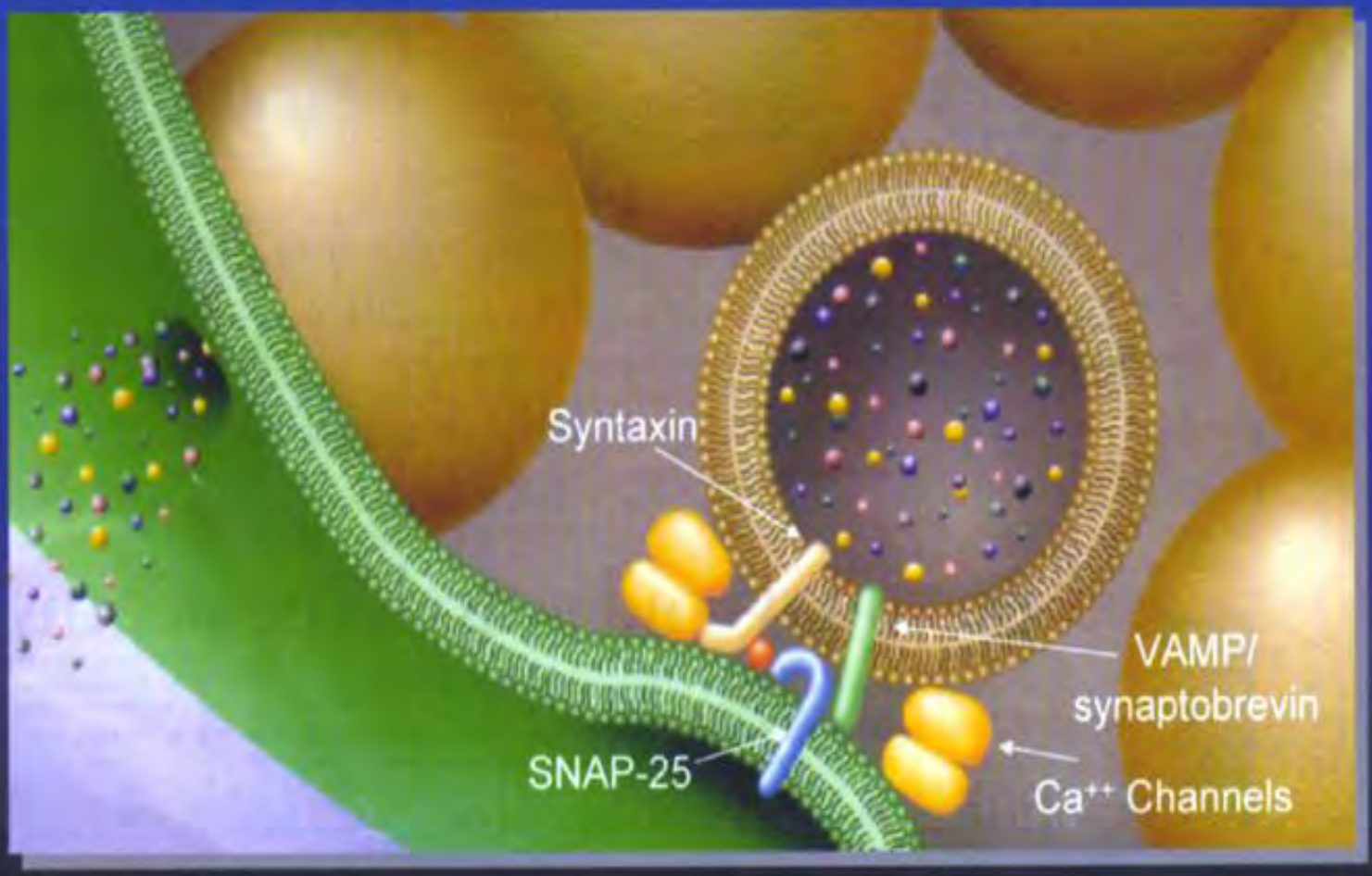
FDA today announced the approval of Botulinum Toxin Type A (Botox Cosmetic) to temporarily improve the appearance of moderate to severe frown lines between the eyebrows (glabellar lines), a medical condition that is not serious. The product's manufacturer, Allergan, Inc., Irvine, California, is now allowed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


Nf
Mi
Gly
SV
BL
Mt
IP

Regulated Exocytosis
Multiple Neurotransmitters / Neuropeptides Released From Vesicle



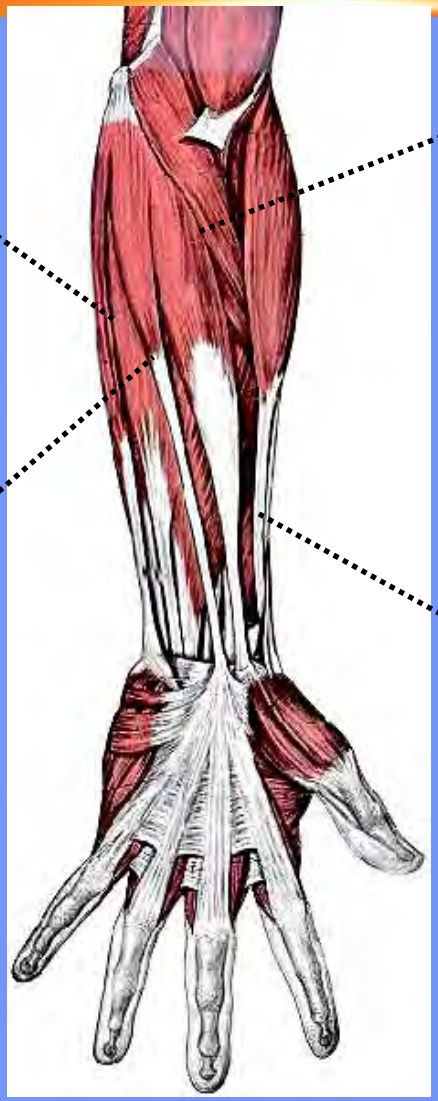
Adapted from *Trends in Cell Biology*, July 1997.

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

Post-Injection
Tibialis Post, Gastroc

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

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

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