Reconsidering the Role of Spasticity in Deformity & Gait Pathology in Children with Diplegic CP

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Reconsidering Spasticity

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Why is “spasticity” such a common focus of concern & intervention in CP & stroke after >20 yrs of research have failed to substantiate its relevance?

What do commonly used “spasticity” assessments reveal about spasticity?

What do passive stretch tests have to do with functional deficits?

Myths About Spasticity

• We understand what spasticity is.
• We know how to detect spasticity in the clinic setting.

“The lack of a precise definition of spasticity may account for the problem of developing a valid, reliable and sensitive method of measurement.”
(Burridge JH, Wood DE et al 2005, p. 69)

“There are no definitive parameters that assess spasticity and there is no universally accepted definition.”
(Seth N, Johnson D et al 2015, p. 109)

The prevailing definition of spasticity...

JW Lance’s (1980):
“a motor disorder characterized by a velocity-dependent increase in tonic stretch reflexes ("muscle tone") with exaggerated phasic reflexes, such as tendon jerks, resulting from hyperexcitability of the stretch reflex, as one component of the upper motor neuron syndrome.”
…is unclear, & has never been validated.
(Pandyan A et al 2005, p. 485)
Problems with terminology within Lance’s definition of spasticity:

- Is hyperreflexia a motor disorder or a somatosensory disorder?
- Is velocity-dependent resistance to stretch specific to or evidence of tonic stretch hyperreflexia? Or of exaggerated phasic reflexes such as tendon jerks?
- Is muscle tone about—or specific to—neurologic, reflex hyperexcitibility?
- Does spasticity require the presence of an UMN lesion? What about spinal cord injury? MS?

Intersecting Issues † Confusion

**THE NEUROLOGIC PROBLEM:**
HYPERREFLEXIA — muscle spindle responses to stretch lack corticospinal modulation.

**THE PHYSIOLOGIC PROBLEM:**
HYPERTONIA - soft tissue transformation increases stiffness (resistance to stretch) after a prolonged history of excessive, tonic use in shortened state.

Spindles are Somatosensory Receptors

- Detect muscle stretch & rate reflexive contraction response.
- Contribute to muscle force generation.
- Are modulated by cortical motor commands at the level of common spinal neurons.

Investigation seeking understanding of the role of these reflexes in functional contexts continues.


Words — and their definitions — drive comprehension and choices among solutions.

*Let’s start with some talk about “tone”...*
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**“Muscle” Extensibility - a.k.a. TONE**

The modulus of resistance – of a group of innervated & relaxed muscle fibers + connective tissue + blood vessels + nerves + skin - to passive stretch.

**Human Resting Muscle Tone (HRMT)**

The passive tonus or tension of skeletal muscle that derives from its intrinsic (EMG-silent) viscoelastic properties; functions inseparably from myofascial tissues & ligaments (& nerves, skin, & vessels).

The vital low-level, passive tension & resistance to stretch that contributes importantly to the efficient maintenance of postural stability in balanced equilibrium positions, magnifying muscle work.


**The Passive Length Tension Curve**

is about Tone in Composite Tissues

**Tone vs. Active Stabilization**

“...co-contraction of muscle is an active neuromotor control [strategy] that provides greater levels of tonus [tissue tension] for increased stabilization.”

(Masi AT, Hannon JC 2008, p. 320)

Lance’s definition pertains to resting muscle tone under passive stretch conditions.
Conclusions:

1) “The nonreflexive resistive torque response to stretch is velocity-sensitive.

2) Both a larger stretch extent [more DF] and muscle initial resistance lead to greater resistive torque increments at high velocity.”

( Lamontagne A, Malouin F, Richards CL. 1997, p. 244)

The Modified Ashworth Scale

The modified Ashworth scale (MAS) is the most widely used method for assessing “spasticity” in clinical practice and research.

The clinician manually stretches the muscle, makes a subjective judgment about the magnitude of resistance (tone) encountered, and assigns a score of 0 (no resistance) to 4 (rigid).

Myth: The Ashworth & Modified Ashworth Scales are Valid Tests of Spasticity

These assessments:

• Actually try to quantify stiffness (tone)

• Use the magnitude of resistance to stretch as evidence of spasticity (hyperreflexia).

• Involve stretching the muscle at one (non-standardized) velocity vs. examining velocity dependence.

• Fail to detect stretch responses (apparent evidence of hyperreflexia) specifically.

The Ashworth & Modified Ashworth Scales

…have been shown to lack strong correlation to EMG & other spasticity measures in these studies:


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MAS Confusion † Wrong Conclusions
A colossal error has occurred in Sweden, where the MAS is thought to quantify spasticity vs. tone. Authors undertook 2,796 examinations in 355 children over 18 years, and concluded:

Ankle DF ROM decreased by a mean of 19° during the first 18 years of life in this population.

“There was a statistically significant association between the ROM and the child’s level of spasticity [MAS score] during the year preceding the ROM measurement.”

(Hägglund G, Wagner P 2011, p. 744.)

A Common, Faulted Assumption
So Hagglund et al (2011) concluded that the MAS score that attempts to quantify tone (stiffness) & cannot detect hyperreflexia predicted that there would be more tone (stiffness) in the future.

Others have made the same mistake:

How About Three Other Spasticity Tests?
• Hoffmann reflex (H-reflex)
• Tendon reflex (T-reflex)
• Stretch Reflex (SR)

Systematic literature review showed:
“Correlations with other (i.e. biomechanical, neurophysiological or clinical) spasticity assessment parameters are moderate to poor. Standardized and broadly accepted protocols are still largely lacking preventing an effective exchange of knowledge.”

(Voerman GE, Gregoric M, Hermens HJ 2005, p. 33)

Lance’s definition produced a firestorm of clarity-seeking & challenging research.
These researchers were unable to confirm the hypotheses that spasticity (hyperreflexia) causes deformity, weakness, loss of dexterity, or gait deficits.

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So, is there another causative factor that escapes the spasticity paradigm?

Let's look at the possible contribution of compromised postural control to deformity development & movement difficulties in children with diplegic CP.

The Drive for Verticality

- Is innate in all living creatures.
- Responds to vestibular, visual, & somatosensory input.
- Preserves life. *(Lie down and stay there, body expects death to follow)*
- Prevails over the kinesiologic correctness of the upright maintenance strategy.

Trunk Control is Associated with Gross Motor Function in Children with CP

Authors found a strong association between segmental trunk postural control and gross motor function and mobility with significant clinical implications for the treatment of children with CP.

*(Curtis DJ, Butler P, Saavedra S et al 2015)*

Trunk control in sitting has a moderate to good correlation with trunk control during gait.

*(Sæther R, Helbostad JL et al 2015)*

Full-term alignment offers a biomechanical assist to postural control (P-C) acquisition.
Biomechanical Shortfalls in Prematurity

Premature neonate

What are they learning?

Essential Ingredients for Postural Control

Antigravity muscle activity develops in a head-to-toe direction & from proximal-to-distal regions.

Extension emerges first. Flexion soon follows.

Bilateral, symmetrical, antigravity extension & flexion are achieved in prone & supine lying by age 4 mos.

(Bly L 2011 & 1994)

#1: Evidence of Antigravity (A-G) Extension

Fundamental Extension Shortfall

LE EXT: Spasticity? Seeking sensory input? Compensation for poor trunk extension?

Ingredient #2: Antigravity Flexion

Flexion Shortfall

Prolonged LE muscle activation: Spasticity? How? Seeking sensory input? Attempt to stabilize to play?
Early Lateral Weight Shifts + Movement
Skull-side shifts displace the COM.
FLX + EXT + unloads face-side limbs + moving!
(Bly L 1994 & 2011)
Ages 5-6 Months

Hypothesis:
Postural control status impacts
lower limb muscle tone & ROM findings
in TD children & in children
with diplegic CP.

Developmental Shortfall
Diplegic CP – Age 26 months
Why no LE use to locomote?

A-G EXT + FLX + P-C in Sitting
7-8 months 8-9 months 9-11 months
Postural Control - Developmental Shortfall
26 months 31 months 26 months

Hamstring Muscle Extensibility in Infancy
Normative Data:
HLT means diminish as the
the length set in utero adapts:
-27° at birth
-18° at 3 months
-10° at 6 months
-1.5° at 9 months
0° at 12 months
(Reade E, et al 1984)

…While balance movement
skills rocket from none to walking!
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**HLT mean, ages 1 to 3 years: -6°**
(Katz K et al 1992)

Between ages 1 ÷ 2.5yrs, R1 end range is absent or negligible, & the HLT finding can be >0°.

Normal hamstring muscle hypotonia.

**The Active Length-Tension Relationship**

Hamstring use history ÷ emergence of a normal 1st catch on the L-T curve = R1 end range. Fascia & muscle adapt to optimize energy use.

R1 end range, a.k.a. “resting” or “functional length,” occurs in a zone in which actin & myosin filaments overlap for optimum contractile force generation.


**Knee EXT Deficit Increases ~ Age 4 Years**

HLT means:
- Girls: -17°
- Boys: -25°

(Katz K et al 1992)

Katz et al did not report R1 & R2 end ranges, but took one measurement at the point of mild resistance (R1)…at which the child experienced pain (R2)…???

**A Common HLT Finding in Children with CP**

Matthew – Age 4 yrs

Diplegic CP

**Body weight is forward & he is still standing…How?**

Ideal R1 at age 4 years = -30°

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Compensatory P-C strategies engage the limbs.

Hypothesis:
If trunk & hip control are inadequately developed, upright maintenance wins over purposeful function.

Limb muscles are recruited tonically & routinely (chronically) to try to gain stability.

Hypothesis: LE Muscles Recruited Tonicly - to Maintain Upright Postures - Shorten

- Gastrocnemius
- Iliopsoas & Rectus femoris
- Hip adductors
- Hamstring muscles

Physiologic Muscle & Fascial Adaptation in CP

Stiff tissues show numerous changes in muscle fiber type (more Tonic type), size, mechanical features, hydration, organization, often with fibrosis.


Physiologic Adaptation to Use History Continues Over Time

- The gastrocnemius muscle volume decreases & stiffness increases in children w/ pyramidal-type CP.
- Other changes:
  - Muscle fiber atrophy
  - "Extracellular Connective tissue (CT) proliferation"
  - Dehydration with muscle volume loss
  - Compromised blood flow

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Myth: Spasticity causes contracture.
Studies of muscle shortening in the presence of chronic, tonic stimulation showed that maintaining tissue length prevented shortening.
Muscle transformation into contracture is myogenic; the process occurs in the muscle & fascia. It is not neurogenic.


Myth: Spasticity causes contracture.
Dystonic, involuntary, chronic, tonic muscle activation or recruitment could lead to contracture formation if the tissues are allowed to shorten.
Dystonia is a motor disorder resulting from a lesion in the basal ganglia.


Evidence that Maintaining Comfortable Tissue Length Can Prevent Deformity in CVA
The R-Wrap© AFO
Made in PF if needed.
Originator: John Russell CPO, FAAOP thermafox@gmail.com

Myth: Spasticity interferes with gait kinematics.
For >20 yrs, studies have indicated that hyperreflexia is of questionable significance in the movement disorder in children & adults with CNS dysfunction.


27 citations, & counting….

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**Confusion Promotes Faulty Practices**

"CATCH": Computer Assisted Technique to Characterize Hypertonia

EMG data obtained in gait.

**Purposes:**
- To integrate measured joint kinematics with muscle EMG data.
- To "quantify spasticity" in a clinic setting & in functional context.

*(Norris JA, Cabrera MN et al 2007)*

...Really?

**Typical Sagittal-Plane Body COM Management in Developing Gait**

Toddlers walk with COM anterior.

Resolution occurs over 7 years.

*(Dierick F, Lefebvre C, et al 2004)*

**Common Strategy - Anterior Body COM**

Many children with CP live in a chronic RR to immature carriage of body COM forward.

Hypothesis: Chronic reliance upon dorsal muscles to remain upright → contractures in those muscles.

**Plasticity ‡ Neuromotor Maps**

Practice – competent or compromised – builds these maps.
Key Resource


The effect on gait of an anterior placement of the whole body center of mass.


Did this TD child become spastic by walking with his COM displaced forward? (Sisson GA et al 1994)

Did this child overcome or cure spasticity by moving his COM backward? (Sisson GA et al 1994)

Children with Diplegic CP Lose DFROM

They typically use the triceps surae muscles tonically and in shortened state to maintain the upright position while the body COM is displaced forward.
Gait pathologies more likely result from:
• Immature or compensatory postural control strategies
• Dysfunctional corticospinal influences on reciprocal inhibition & selective motor control
• Physiologic (nonreflexive) changes in extensibility in the involved muscles & connective tissues (pathologic stiffness)
• Weakness
• Compromised vestibular & somatosensory inputs
• Dystonia
• Ataxia.

Myth: The Cortical Lesion
Spastic cerebral palsy is often associated with damage to or developmental differences in the part of the brain called the cerebral cortex.

www.gillettechildrens.org/conditions-and-care/cerebral-palsy/

…Really?

Diffusion Tenser Imaging (DTI) (or dMRI)
A form of magnetic resonance imaging that allows in vivo examination of the presence & integrity of white matter tracts via water diffusion; the current method of choice for studying white matter pathology & reorganization.


News from DTI about CP S/P PVL
• No DTI studies report a static UMN lesion.
• Descending corticospinal tracts can resemble those of TD peers.
• Faulty tract formation is often evident in white matter fibers over many regions of the brain, & particularly those connected to the ascending sensory cortex.

Children with CP show significant somatosensory deficits & inefficient cortical processing of input, as determined via clinical tests of PPC & tactile sensation, & via MRI, fMRI, SEPs, VEPs, DTI, & MEG.

So, what causes CP?

A stronger, more current definition:

“Current research suggests the majority of Cerebral Palsy cases result from abnormal brain development or brain injury prior to birth or during labor and delivery.”

http://www.cerebralpalsy.org/about-cerebral-palsy/definition

“Half of being smart is knowing what you’re dumb at.”

- Soloman Short

We certainly don’t have this one nailed…

What do we need to know more about…?

Fascial Adaptation - An Emerging Science

“Recent” developments in research on global fascial distribution, architecture, & operational mechanisms reveal adaptive responses to prolonged tension that might be mistaken for spasticity.
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New Revelations from Video Images

Fascia surrounds, penetrates, & connects skeletal muscle, joints, organs, nerves, vascular beds, & skin throughout the body.

In skeletal muscle, the lattice-like architecture is open & wet, holding most of the water in the body; elastic tubules support & accommodate movements by sliding along, detaching, & reattaching with each other.

(Fuimberteau JC 2012)

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Fabulous Fascia Videos

- https://www.youtube.com/watch?v=uzY8-wQzQMY
- https://www.youtube.com/watch?v=qSXpX4wy0Y8
- https://www.youtube.com/watch?v=uxQSn87Hq0

Thomas Myers – Anatomy Trains:
https://www.youtube.com/watch?v=uxQSn87Hq0

Strolling Under the Skin – JC Guimberteau
https://www.youtube.com/watch?v=eW0lvOVKdx

Schleip lecture: using manual Rx to influence water flow vs. affect collagen cross-links - much slower & more gentle.
https://www.youtube.com/watch?v=y9NqWZ-0ik

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Fibroblasts Regulate Fascial Status

The fascial network contains fibroblasts (a.k.a. fibrocytes):
- Highly responsive to mechanical stimuli – particularly tension
- Play critical roles in producing & regulating the integrity of extracellular matrices & tissue fluid volume & pressure, & in wound healing.


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Fibroblast Behavior Under Stretch

After long-term mechanical stress (days to weeks) and/or injury, fibroblasts respond by increasing a-actin synthesis & transforming into contractile myofibroblasts that produce ECM with adhesion sites.

Myofibroblasts close wounds & actively protect tissues from separating.

(Myofibroblasts close wounds & actively protect tissues from separating.

(Langevin HM, Storch KN et al 2006)

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Do myofibroblasts (MFB) contribute to the clinical impressions of spasticity?

“When fascia contracts it is capable of stimulating muscle stretch receptors (muscle spindles)...”

http://tuckeypt.com/for-the-viscera/

- Do fibroblasts in CT sense excessive tension in shortened muscles & differentiate into MFB?
- Since MFB also produce smooth muscle actin that is contractile, is this contractility perceived in a “catch” or quick-stretch-induced contraction?

Nerve Entrapment in CP

29 patients with CP and severe deformities
Nerve conduction & needle EMG
- Nerve conduction abnormalities were detected in 32 of 400 sensory or motor nerves tested.
- 11 patients (37.9%) showed abnormal nerve conduction indicating entrapment neuropathy.
- Contractures & deformities can cause nerve damage, possibly as a result of the stretching, angulation, or compression mechanisms in the anatomic fibro-osseous passages, where nerves are particularly susceptible.

(Frascarelli M, Frascarelli F et al 2005)
If spasticity causes movement disorders & deformities, then treating “spasticity” should cure them.

(Lin JP 2011, Pandayan AD et al 2005; Sheean GL 2001)

Reconsidering Spasticity as a Treatment Target

After failing to find strong correlations between midrange MAS scores & EMG output in children with CP, Damiano et al (2002) concluded:

“Patients assigned Ashworth scores in the mid-ranges of resistance yet who had passive stiffness alone could be incorrectly judged to have mild to moderate spasticity and, consequently, may be treated inappropriately.”

(Damiano DL, Quinlivan JM et al 2002, p. 117)

Lorentzen et al (2012) note that spasticity is a frequently used diagnosis, provoking widespread use of anti-spastic medication. A review of the neuroscience lead them to conclude:

“...it is necessary to develop better tools for the clinical diagnosis of spasticity in order to avoid potential malpractice and to limit treatment with anti-spastic drugs to patients with documented increased reflex-mediated muscle tone as their main annoyance.”


Reconsidering Spasticity as a Treatment Target

With or without spasticity, address postural alignment & control deficits:

• Optimize joint alignment & weight distribution through the torso, LE joints, & feet.
• Correct & enhance PPC & cutaneous sensory input.
• Work to eliminate the necessity to recruit limb muscles all day long to maintain upright postures.
• Building essential components of balance & movement requires massed practice.
So why is Botulinum toxin-A considered a “best practice” in CP care?

Maybe because 53.6% of the 28 industry-sponsored studies of BTX effectiveness had favorable conclusions unlike only 20% of the 25 non-industry-sponsored studies?

The efficacy of using BTX-A injections in CP palsy is controversial. (Sung Sung KH, Chung CY et al 2013)

BTX-A is anti-activation = paralytic.

BTX-A & its derivatives act to denervate the muscle by inhibiting the release of acetylcholine at the neuromuscular junction.

Systematic, long-term data on the physiology, morphology, & recovery of human muscles exposed to BTX-A treatments are not available.

The US FDA has never approved this off-label use. http://www.fda.gov/NewsEvents/Newsroom/PressAnnouncements/ucm149574.htm

Systematic, long-term data on the physiology, morphology, & recovery of human muscles exposed to BTX-A treatments are not available.

BTX-A injections into the gastrocnemius muscles to “reduce spasticity”:

a) Do not prevent equinus deformity (Tedroff K et al 2009)
b) Might not do anything (Ackman JD et al 2005)
c) Do not contribute to the success of BK casting (Ganzman AM et al 2004)
d) Do not enhance the effects of casting in the long term (Kay R, Rethelfsen SA, et al 2004)

Research has shown that BTX-A injections:

f) Have resulted in atrophy, fatty tissue and CT infiltration, muscle fiber loss, & degradation of injected and non-injected muscle in animals. (Fortuna R et al 2011 & 2015)
g) Do not improve mapping (DTI) or GMFM scores compared with PT alone after 6 months. (Chaturvedi SK, Rai Y, et al 2013)
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Summary

• No one has proven that velocity-dependent resistance to passive stretch is clear evidence of the presence of a poorly-modulated reflex response to stretch.
• Spasticity (if it is indeed hyperreflexia) cannot cause deformity. It is a reaction, not an action.
• Chronic, tonic muscle use – either dystonic or 2º postural control deficit - can cause deformity.
• Muscle transformation is myogenic vs. neurogenic.
• The MAS is a test of resistance to stretch - so, of muscle tone - vs. spasticity.

Summary

Velocity-dependent resistance to stretch might be caused by:
• A component of normal, nonreflexive, viscoelastic muscle & fascial tissue properties
• Nerves and blood vessels bound by adhesion & placed under stretch
• Dehydration-related stiffness accompanying muscle atrophy & fascial shortening
• The activity of myofibroblasts that might be present after a prolonged state of tissue tension.

Summary

• Spasticity – if present – is of questionable significance in gait pathology compared with corticospinal dysfunction.
• Spasticity, if present, is a somatosensory system deficit. Optimize somatosensory input.
• The evidence reveals that the common focus on spasticity - as a cause of pathology & a target for treatment - is misguided in the CP population.
• “Spasticity” treatments can increase ROM by weakening muscles, but they do not produce postural control, strengthen muscles, or normalize functional strategies.

Thank you the honor and the opportunity to share these issues with you.

Questions…?

…Discussion?
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ACPOC Presentation – April 14, 2016 - References and Readings

On Spasticity - Assessment, Management, Influence on Contracture


Damiano DL, Quinlivan JM, Owen BF, et al. 2002. What does the Ashworth scale really measure and are instrumented measures more valid and precise? Dev Med Child Neurol. 44(2): 112-118.


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Spasticity - Influences on Gait? - Pediatrics


Spasticity - Influences on Gait? - Adults


**Muscle Physiology, Kinesiology, & Pathophysiology**


**Postural Control**

Bly L. 2011. Components of Typical and Atypical Motor Development. Laguna Beach, CA; Neurodevelopmental Treatment Association; www.ndta.org


**Fascia**


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**Peripheral Nerve Constriction in “Spastic” Muscle**


**Brain Imaging**


