Evaluation Skills Part 2: Hypotonia

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Objectives

- The learner will be able to explain the definition of Hypotonia and how to assess muscle tone with the pediatric patient.
- The learner will be able to differentiate the common causes of Hypotonia in the pediatric patient.
- The learner will be able to perform an evaluation of a child with Hypotonia.
- The learner will develop a personal “resource toolbox” to assist in on-going treatment of patients with Hypotonia.
What is muscle tone?

- Muscle tone is assessed by resistance to passive stretch.
- 2 types of tone:
  - Postural: Passive resistance to movement of axial muscles: neck, back, trunk
  - Phasic: Passive resistance to movement of extremities: appendicular structures
Assessing Muscle Tone

- Postural tone:
  - In a younger infant: ventral suspension
  - In an older infant: position of trunk in sitting and standing
Assessing Muscle Tone: Phasic (extremity) tone

- Phasic (extremity) tone:
  - Scarf sign
    - Normal = elbow between the bilateral midclavicular lines
    - Low = elbow crosses the midline to contralateral midclavicular line
    - High = elbow does not cross the ipsilateral midclavicular line
Assessing Muscle Tone

- Popliteal angle
  (Hamstring length test)
  - Normal = 6 degrees 1-3 yo; 17-27 degrees in 4 yo; 26 degrees in > 5 yo
  - Low = < 6 degrees in 1 yo
  - High = ≥ 50 degrees in ≥ 1 yo
What is Hypotonia

- Muscles are slow to initiate a muscle contraction, contract very slowly in response to a stimulus and cannot maintain a contraction for as long as a “normal” muscle
- Muscle tone is regulated by signals that travel from the brain to the nerves and tell the muscles to contract. (website)
- Is a symptom of an underlying disorder
- May result from a change in the properties of the peripheral structures such as muscles and tendons as well as from changes in reflex pathways
- Hypotonia can happen from damage to the brain, spinal cord, nerves, or muscles. The damage can be the result of trauma, environmental factors, or genetic, muscle, or central nervous system disorders. (website)
Hypotonia

- “Weakness” and “hypotonia” are 2 different things
- Weakness: inability of a muscle to generate sufficient tension in a muscle for postural control and active movement
- Hypotonia comes from muscle itself and recruitment of motor units and timing of their activation
- Can increase muscle strength
Clinical characteristics of Hypotonia

- Decreased strength
  - Decreased tonic contraction against gravity

- Hypermobile joints
  - Poor stability when weightbearing

- Floppy
  - Decreased state of readiness for movement

- Needs external supports
  - Limited weightbearing
  - Can’t sustain postural control against gravity
Clinical characteristics of Hypotonia

- Feeding, respiratory difficulties
- Delayed motor skills
  - Poor head control
  - Abnormal movement patterns
  - Impaired mobility
- Decreased activity tolerance
Etiologies of Hypotonia

- Central Nervous System
- Peripheral Nervous System
- Muscle itself
- Neuromuscular Junction
- Hypotonia due to hypofunctioning vestibular system
Vast majority of infants who present before age one with hypotonia have a CNS etiology

Hypotonia of CNS origin accounts for 66-88% of cases

- Genetic chromosomal disorders >31%
- Structural brain anomalies >13%
- Myopathies >5%
- Spinal muscular atrophy >2%
- Muscular dystrophy > 2%
CNS Hypotonia

- Benign Congenital Hypotonia
  - Hypotonia at birth
  - Persisting motor coordination difficulties
  - Tone improves over time
  - Good strength and DTRs
- Chromosomal Disorders
  - Prader-Willi
  - Angelman Syndrome
  - Down Syndrome
- Brain Malformation
  - Lissencephaly
  - Pachgyria
  - Septo-optic Dysplasia
  - Agensis of Corpus Callosum
CNS Hypotonia

- Brain injury
  - Hypoxic-ischemic encephalopathy
  - Intracranial hemorrhage
  - CNS infection
  - Trauma
  - Hydrocephalus

- Neurometabolic Disorders
  - Peroxisomal
  - Myelin Disorders
  - Mitochondrial
  - Amino and Organic Acidurias

- Spinal Cord Dysfunction
  - Insults
  - Trauma
  - Tumor
CNS Hypotonia Characteristics

- Cannot sustain postural control
  - Lack of weightbearing
- Delayed motor milestones
- Intact DTRs
- Associated with Dysmorphic features
- Motor coordination difficulty

- Have adequate strength
  - Show anti-gravity movement but cannot sustain
- Often with Additional deficits
  - Behavior issues
  - Cognitive deficits
  - Impaired neurological function
  - Spasticity
PNS Hypotonia

- Spinal Muscular Atrophy
  - Hypotonia at birth
  - Proximal and distal weakness
  - Absent DTRs
  - EMG: abnormal fasciculations
- Poliomyelitis
  - Asymmetric, flaccid paralysis
  - EMG: abnormal fasciculations
  - Decreased or absent DTRs

- Peripheral Neuropathy
  - Congenital hypomyelinating neuropathy
  - Mitochondrial disorder
  - Demyelinating neuropathy
  - Charcot-Marie Tooth
PNS Hypotonia Characteristics

- Muscle Fasciculations
- Delayed Motor Milestones
- Decreased or Absent DTRs
- Muscle Atrophy
- Impaired Strength
- Abnormality Usually Limited to Muscular System
CNS vs PNS

Central
- Some or adequate strength
- Some anti-gravity movements
- Decreased or increased DTRs
- Slow placing reactions
- Has motor and cognitive delays
- Head circumference may be smaller than normal

Peripheral
- Poor to no strength
- No anti-gravity movements
- No DTRs
- No placing reactions
- Has motor delays but not cognitive
- Fasciculations
- Joint contractures possible
Neuromuscular Junction and Muscle Itself

- Disorders of Myoneural Junction
  - Myasthenic syndromes
  - Infant botulism

- Muscle disorders
  - Metabolic myopathy
  - Congenital myopathies
  - Muscular dystrophies (Gower sign)
Hypofunctioning vestibular system

- Bodensteiner, et al (2003) found:
  - 4 children out of 31 in their study had hypotonia and hearing loss
  - Children with this type of hypotonia tend to outgrow hypotonia as they age
  - Suggest: inclusion of hearing assessment in all children hypotonic enough to warrant investigation
  - Do CT with thin cuts of the temporal bone to look at vestibular apparatus (More reliable than MRI)
Assessing Low Muscle tone

- Subjective information
  - Typical schedule (how much time spent lying flat on back or stomach)
  - Position for feeding
  - Thyroid function
  - Medications
  - Medical complications
  - General arousal and movement of the child
  - Sleep pattern
  - Atlanto-axial instability
  - Vision concerns
Assessing Low Muscle Tone

- Objective
  - Infant Neurological International Battery (INFANIB)
  - Movement Assessment of Children (MAC)
  - Gross Motor Function Measure (GMFM)
  - PIQ
  - Developmental testing
    - PDMS-II
    - BOT
Assessing Low Muscle Tone

- INFANIB
  - Scarf sign
  - Heel-to-ear
  - Popliteal angle
  - Leg abduction

- These items have low reliability

- Question to whether INFANIB assesses muscle tone, muscle extensibility, or ligamentous laxity
Scarf Sign

- 0-3 months = 0-15 degrees
- 3-6 months = 15-45 degrees
- 6-9 months = 45-60 degrees
- 9-18 months = 60-85 degrees
Popliteal angle

- 0 to 3 months = 80-90 degrees
- 3-6 months = 90-110 degrees
- 6-9 months = 110-150 degrees
- 9-18 months = 150-170 degrees
Hip Abduction

- 0-3 months = 80-90 degrees
- 3-6 months = 90-110 degrees
- 6-9 months = 110-150 degrees
- 9-18 months = 150-170 degrees
Assessing Low Muscle Tone

- MAC
  - Looks at fine and gross motor control
  - Can be done in 30 minutes or less with observation only; don’t have to handle the child
  - Normed in Denver from 2006-2007
  - For children from 2-24 months
  - Evaluates
    - General Observations (behavior state; autonomic NS stability)
    - Special Senses (visual tracking, peripheral vision, hearing)
    - Head Control
    - UEs and hands
    - Pelvis and LEs
    - Primitive reactions
    - Muscle tone
    - Has L and R side scores
Assessing Low Muscle Tone

- GMFM
  - Hard to do all 88 items for older children, as they don’t want to do younger items
  - Since it’s criterion-referenced the children with DS do not have to be compared with “norms”
  - More responsive to motor change over time with children with DS than the BSIDII.
Assessing Low Muscle Tone

- PIQ
  - Posture in Positions
  - Initiation and Inhibition
  - Quality

- Positions
  - Supine
  - Prone
  - Sitting
  - Standing
  - Quadruped
  - Kneeling
Supine

- Posture in Positions
  - Look for thoracic kyphosis and flattening of rib cage with more horizontal orientation of ribs
  - Frog leg position in supine
  - Ability to maintain midline position of head

- Initiation and Inhibition
  - Do they reciprocally move LEs/UEs against gravity
  - Can they bring hands to midline
  - Do they initiate rolling supine to prone
  - Can they track toys visually
  - Can they transition from supine to sitting
  - May be inhibited from getting chin tuck as increased cervical extension overlengthens cervical flexors

- Quality
  - Do they roll with trunk dissociation or log rolling
  - Do they transition to sitting with trunk rotation pushing up from ground or through sagittal plane only
  - Head lag during pull to sit
Supine
Prone

- Posture in Positions
  - Use cervical hyperextension to lift head in prone then it rests on occiput (Cervical stacking)
  - Uses eyes to lift and hold head up; May use tongue retraction to hold head up
  - Frog leg position in prone
  - Hyperextension of elbows when pushing up in prone

- Initiation and Inhibition
  - Push up through hands in prone
  - Reach for toys in prone
  - Do they initiate rolling prone to supine
  - Do they transition from prone to sitting
  - May initiate movement with phasic bursts of muscles (lifting head with cervical extension)

- Quality
  - Dissociation of trunk when rolling prone to supine
  - Weight shifting when reaching for toys in prone
  - Prone to sitting through straight plane movements with minimal rotation
Prone
Sitting

- Posture in Positions
  - How do they prefer to sit: W-sitting; Long sitting; Criss-cross sitting; etc
  - Paraspinal weakness results in total kyphotic posture with posterior pelvic tilt and forward head posture (Limits anterior movement preventing normal pressures)
  - Hip IR/ER; Look at foot position

- Initiation and Inhibition
  - Reach for toys on the ground or held off the ground
  - Crossing midline in sitting
  - Transition between sitting and prone/quadruped

- Quality
  - Transition over hip or with hips in abduction
  - Place hand down to reach outside BOS
  - Transfer toy between hands rather than crossing midline
Sitting
Standing

- Posture in Positions
  - Children assume WBOS and low COM: muscles placed in poor length-tension position
  - Bony deformity of forefoot
  - Pronated flat foot
  - Calcaneal valgus
  - Knee valgus
  - Hip IR/ER
- Initiation and Inhibition
  - Squatting and returning to standing
  - Reaching in all planes without LOB
  - Raise on tiptoes to reach for toys overhead
- Quality
  - Ankle/hip strategy
  - Knee alignment when squatting
  - Push-off on metatarsals when raising on tiptoes
Standing
Quadruped

- Posture in Positions
  - Elbow hyperextension
  - Lumbar lordosis
  - Cervical stacking when lifting head
  - Hip abduction/ER

- Initiation and Inhibition
  - Transitioning between sitting and quadruped
  - Rocking in quadruped
  - Reaching in quadruped
  - Creeping over even terrain
  - Creeping over small obstacles
  - May initiate movement with phasic bursts of muscles (lifting head with cervical extension)

- Quality
  - Weight shift when creeping
  - Transition to sitting with increased hip abduction
  - Endurance for creeping
Tall Kneeling/Half Kneeling

- Posture in Positions
  - Hanging on ligaments for balance
  - Lordosis with decreased core activation
  - Excess hip abduction/ER for widening BOS
  - Poor ankle stability in half kneel
  - Resting on heels

- Initiation and Inhibition
  - Transitioning from tall kneel to half kneel
  - Pulling to stand at a support surface
  - Transitioning to standing without support surface
  - Reach and rotate head without LOB

- Quality
  - Weight shifting for transitioning tall kneel to half kneel
  - Excess use of UEs to pull to stand
  - Bear crawl to stand without support surface
Kneeling
Assessing Low Muscle Tone

- **ROM**
  - Check for tight pecs, lats
  - Tight intercostal muscles

- **Functional Strength testing**
  - Bridges
  - Squatting
  - Floor to stand transitions
  - Sit to stand transitions
  - Jumping/Hopping on 1 foot
  - Stair navigation
Assessing Low Muscle Tone

- **Static Balance**
  - SLS with EO and EC
  - Tandem stance with EO and EC
  - Balance on tiptoes

- **Dynamic Balance**
  - Tandem walking on balance beam
  - Walking backwards and laterally
  - Stairs without handrail
Assessing Low Muscle Tone

- Sensation and Perception
  - Depth perception may be compromised in children with hypotonia; its development relies on convergence of the eye and self-initiated movement through space
    - Tracking and eye-hand coordination
    - Up and down stairs: depth perception may be an issue
    - Increased incidence of cataracts and strabismus
  - Use eyes to hold head up = decreased visual exploration
  - Children with DS have increased incidence of hearing problems or may have increased sensitivity to sound
Assessing Low Muscle Tone

- Proprioceptors
  - Sensors in limbs that give information about joint angle, muscle length and tension = give information about where limb is in space
  - Golgi tendon organs, muscle spindles
  - Proprioception from Latin proprius, meaning “one’s own” and perception, is the sense of the relative position of neighboring parts of the body and strength of effort being employed in movement.
Assessing Low Muscle Tone

- **Testing Proprioceptors**
  - **Limb movement sense test**
    - Child keeps eyes closed while you passively move one limb; imitate movement with opposite limb. Do in chair for UEs, supine for LEs
    - Score as either can do or cannot
  - **Limb-matching test**
    - Child’s eye closed; you position one extremity and they must duplicate position
    - Score
      - 2 = no error in matched position
      - 1 = position approximately correct; joint angles incorrect up to 30 degrees total
      - 0 = one or more joint angles in error by > 30 degrees
  - **PDMS-II Imitation of movement**
    - Have child imitate movements when UEs are out of visual field
Intervention Strategies for Children with Low Muscle Tone

- Rolling
- Independent sitting with good postural alignment
- Independent standing with good skeletal alignment
- Self-initiation of cruising and motivation to move
- Single limb stance
- Transitioning from the ground to standing
- Jumping forwards and down
Rolling

- Impairments
  - Decreased core strength with elevated rib cage and decreased oblique muscle activation
  - Decreased cross-plane movements and trunk dissociation with minimal rotation
  - Impaired visual tracking
  - Decreased strength of hip flexors for initiating rolling supine to prone
  - Decreased cervical spine strength for clearing head
  - Decreased tolerance for tummy time
  - Decreased UE strength for pushing up through arms
  - Impaired motor planning and coordination

- Treatment Ideas
  - Kinesiotaping to activate obliques
  - Transitioning supine to sitting through sidelying overground and on stability ball for oblique activation
  - Lateral head righting for clearing head against gravity when rolling
  - Active and passive trunk rotation and dissociation
  - Reaching for knees/feet in supine
  - Prone positioning and reaching on stability ball, overground, and in carrying hold
  - Reaching for toys in supine and across body
Kinesiotaping obliques
Independent sitting with good postural alignment

- **Impairments**
  - Decreased muscle strength and endurance in postural muscles
  - Decreased muscle tone in trunk and extremities
  - Increased joint mobility and ligamental laxity
  - Vision deficits
  - Delayed righting and protective reactions

- **Treatment Ideas**
  - Sitting in Bumbo with towel folded to promote neutral pelvic alignment
  - Adaptive equipment: Theratog, kinesiotaping, SPIO for promoting upright posture
  - Manual cuing to promote upright posture and pelvic alignment when sitting
  - Propped sitting at a bench for decreasing posterior pelvic tilt
  - Sit on ball: work trunk flexors and extensors
  - Pivoting in prone for weight shifting and oblique strengthening
  - Head and trunk righting
  - Rolling supine to/from prone for core and UE strengthening
Adaptive Equipment

- Stabilizing Pressure Input Orthosis (SPIO)
- Theratog
- Kinesiotaping
  - Activating rectus abdominis
  - Activating back extensors
Adaptive Equipment

- SPIO
  - Level Four: Conover NC
    - (828) 328-5347
  - Griffin Home Healthcare: Charlotte NC
    - (704) 347-1993 Contact: Richie Griffin
  - Center for O&P Care: Durham NC
    - (919) 797-1232
  - Skyland P&O: Fletcher NC
    - (828) 684-1644 Contact: Angela Polson
  - Hanger Clinic: Wilmington NC
    - (910) 791-3333 Contact: Matthew Murphy
  - Hanger Clinic: Whiteville NC
    - (910) 640-1211 Contact: Matthew Murphy
  - Hanger Clinic: Charlotte NC
    - (704) 377-7099
Independent standing with good skeletal alignment

- **Impairments**
  - Decreased muscle strength and endurance
  - Decreased muscle tone in trunk and extremities
  - Increased joint flexibility
  - Poor static balance: Hypofunctioning vestibular, vision, or somatosensory systems
  - Poor proprioceptive sense
  - Poor tactile input in foot

- **Treatment Ideas**
  - Orthotic
  - Sit on ball: work trunk flexors and extensors
  - NMES/FES to gastrocs, glute max, abdominals
  - Theratog and kinesiotape to align scapula and decrease rounded shoulders
  - Rows/Tug of war for mid scapular muscle activation
  - Side bridges, lateral step ups, clam shells for glute med strengthening
  - Work glute max and lats simultaneously for increasing core stability
  - Practice equilibrium strategies and uprtrnain the weak system
  - Ankle weights for increasing proprioception
  - Picking stickers off feet with and without vision
  - Bubble wrap to prevent hyperextension
Adaptive Equipment

  - 4 clinical criteria for flexible flat foot
    - Calcaneus rests in a valgus position
    - Medial protrusion of the head of the talus over the medial aspect of the foot upon palpation
    - Flattened longitudinal arches
    - Inversion of the heel and restoration of medial arch when the great toe was passively extended
  - Also noted:
    - Stood with excessive medial WBing such that a piece of paper slid underneath her lateral calcaneus and fifth ray no loading occurred on the paper
    - WBing on navicular head
    - Tibial internal rotation
    - Walked with excessive pronation, tibial internal rotation, knee recurvatum, and excessive lumbar lordosis
  - Fabricated in-shoes orthotics to address these issue and falling decreased and scores on PDMS-II increased into average range
Adaptive Equipment

SMOs

Little Steps

Pattibobs
Little Steps in Action
Self-initiation of cruising and motivation to move

- Impairments
  - Poor cognitive skills
  - Poor motor planning (may look like weakness)
  - Decreased vision, hearing, or touch
  - Poor LE dissociation and coordination
  - Decreased muscle strength of hip muscles
  - Poor stability of pelvis: Decreased strength of pelvic muscles
  - Decreased lateral weight shifting

- Treatment ideas
  - Pair 2 senses (vision and auditory)
  - Give hands-on help to move through sequence and then decrease assistance
  - Facilitation to prevent compensation of hip flexors
  - Hitting ball with side of foot in standing
  - Zoom ball with 1 foot abducted
  - Play in tall kneel and half kneel (on/off compliant surface)
  - Twinkle little star with lateral weight shifting
  - Lateral stepping up/down a bench
  - Sit to/from stand transitions with facilitation to maintain hips in neutral
SLS

- Impairments
  - Poor stability of pelvis
  - Decreased strength of pelvic muscles (hip and abdominal)
  - Increased joint flexibility
  - Poor static balance: Hypofunctioning vestibular, vision, or somatosensory systems
  - Poor proprioceptive sense
  - Poor tactile input in foot

- Treatment Ideas
  - Step on/over phone book as part of cruising
  - Stand with one shoe on, other off
  - Play in tall kneel and half kneel (on/off compliant surface)
  - Side bridges, lateral step ups, clam shells for glute med strengthening
  - Work glute max and lats simultaneously for increasing core stability
  - Don shoes/socks in standing
  - Strengthen ankle
Transitioning from the ground to standing

- **Impairments**
  - Poor stability of pelvis
  - Decreased strength of pelvic and hip muscles
  - Decreased cross-plane movements
  - Increased joint mobility and ligamental laxity
  - Poor motor planning
  - Impaired weight shifting

- **Treatment Ideas**
  - Step on/over phone book as part of cruising
  - Play in tall kneel and half kneel (on/off compliant surface)
  - Side bridges, lateral step ups, clam shells for glute med strengthening
  - Bridges, squats, and lunges for glute strengthening
  - LAQs, lunges, wall squats for quad strengthening
  - Strengthen ankle
  - Stand through bear crawl initially and work towards standing through half kneel
Floor to stand transition.MOV
Jumping Forwards and Down

- Impairments
  - Decreased concentric strength in hip extensors, quads, and plantar flexors
  - Decreased eccentric hip extensor and quad strength during landing
  - Decreased power generation
  - Hypermobility of joints
  - Poor motor planning and coordination

- Treatment Ideas
  - Jumping into deep squat to prevent genu recurvatum
  - Squat jumps for increasing power generation
  - Rapid sit to stands for increasing power generation paired with slow stand to sit transitions for improving eccentric control
  - Squatting and raising on tiptoes for improving motor planning
  - Facilitation for push-off when raising on tiptoes
  - Running for increasing power in gastrocs for push-off
  - Hip abductor strengthening for decreasing stress on medial knee during landing
Adaptive Equipment

Before

After

Hip Helpers:
Limit excessive hip abduction and maintain hips in neutral alignment

Stander

Gait trainer

Hand Grips
Forearm Platform (pair)
Take-aways for Hypotonia

- Decreased rotation and movement in transverse plane
  - Obliques do not fire to stabilize rib cage
- Poor endurance for activity
  - Elevated ribcage does not allow for full expansion of diaphragm
- Decreased weight shifting during transitional movements
- Hypermobile joints with ligamental laxity
- Poor postural control results in needs for orthotics and adaptive equipment
- Poor balance
  - Decreased proprioceptive input through joints and somatosensory input
- Poor postural endurance results in decreased attention for fine motor activity
- Decreased Arousal
- Visual impairments
  - Laxity in eye muscles
  - Impaired depth perception
  - Decreased visual scanning
QUESTIONS?