Evaluation Skills: Hypotonia – Part 1

Muscle tone is assessed by resistance to passive stretch. There are 2 types of tone:

- Postural: Passive resistance to movement of axial muscles: neck, back, trunk
- Phasic: Passive resistance to movement of extremities: appendicular structure

How to assess the postural tone of a patient depends on the age. In a younger infant ventral suspension is used. In an older infant it's the position of the trunk when sitting on a bench with no foot support.

How to assess phasic tone depends on if you are assessing the upper or lower extremities. For the upper extremities you can use 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

For the lower extremities you can use the popliteal angle.

- Normal = 6° for 1-3 years old; 17-27° for 4 years old; 26° in 5 years and older
- Low = < 6° in 1 year old
- High = > 50° in > 1 years old

With Hypotonia, muscles are slow to initiate a muscle contraction and contract very slowly in response to a stimulus. Muscles with Hypotonia also cannot maintain a contraction for as long as a "normal" muscle.

Hypotonia is usually a symptom of an underlying disorder. It 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.

Weakness and Hypotonia are 2 different things although weakness is commonly seen with Hypotonia. Weakness is the inability of a muscle to generate sufficient tension in a muscle for postural control and active movement, while Hypotonia comes from muscle itself and recruitment of motor units and timing of their activation.

Some of the clinical characteristics of Hypotonia are decreased strength, hypermobility of the joints with increased flexibility, decreased state of readiness (aka "floppiness"), the need for external support, feeding and respiratory difficulties, delayed motor skills, and decreased activity tolerance.

There are many different causes of Hypotonia. It can come from the Central Nervous System, Peripheral Nervous System, the muscle itself, the Neuromuscular Junction, or it can be due to a hypofunctioning vestibular system.

Hypotonia of the Central Nervous System (CNS) is the most common and accounts for 66-88% of cases. Within CNS Hypotonia are genetic chromosomal disorders (over 31%), structural brain anomalies, myopathies, spinal muscular atrophy, and muscular dystrophy.

Causes represented within CNS Hypotonia is Benign Congenital Hypotonia (Hypotonia at birth), chromosomal disorders, brain malformation, brain injury, neurometabolic disorders, as well as spinal cord dysfunction. The major characteristics of CNS Hypotonia are lack of ability to sustain postural control, lack of weight bearing, delayed motor milestones, intact DTRs, associated with dysmorphic features, and motor coordination difficulty. Patients will display adequate strength, but lack endurance. It is often seen with additional deficits such as behavior issues, cognitive deficits, impaired neurological function, and spasticity.

Causes represented within Peripheral Nervous System (PNS) Hypotonia is Spinal Muscular Atrophy, Poliomyelitis, and Peripheral Neuropathy. The major characteristics of PNS Hypotonia are muscle fasciculations, delayed motor milestones, decreased or absent DTRs, muscle atrophy, impaired strength, and / or abnormality usually limited to muscular system.

CNS Hypotonia vs. PNS Hypotonia

Central	**Peripheral**
Some or adequate strength	Poor to no strength
Some anti-gravity movements	No anti-gravity movements
Decreased or increased DTRs	No DTRs
Slow placing reactions	No placing reactions
Has motor and cognitive delays	Has motor delays but not cognitive
Head circumference may be smaller than normal	Fasciculations & Joint contractures possible

Causes represented by Hypotonia of Neuromuscular Junction or muscle itself are Myasthenic syndromes, infant botulism, metabolic myopathy, congenital myopathies, and muscular dystrophies.

The last causes can be Hypotonia of a hypofunctioning vestibular system. New research suggests that children with a hypofunctioning vestibular system are at a risk for hearing loss, and should therefore have a hearing assessment.

The first place to start in assessing a patient with Hypotonia is gathering the subjective information to include typical schedule (how much time spent lying flat on back or stomach or container device), position for feeding, medications, medical complications, general arousal and movement of the child, sleep pattern, atlanto-axial instability, and vision concerns.

Some objective tests that are generally recommended specifically for Hypotonia are Infant Neurological International Battery (INFANIB), Movement Assessment of Children (MAC), Gross Motor Function Measure (GMFM), and developmental tests such as the PDMS-II or the BOT.

INFANIB is a quick screening using scarf sign, heel-to-ear, popliteal angle, and leg abduction, but these items can have a low reliability.

MAC looks at fine and gross motor control, and can be done in 30 minutes or less with observation only; don't have to handle the child. This is great for children who don't readily warm up. This is for children from 2-24 months. It evaluates general observations (behavior state; autonomic NS stability), special senses (visual tracking, peripheral vision, and hearing), head control, UEs and hands, pelvis and LEs, primitive reactions, and muscle tone. It also has left and right side scores.

GMFM is more associated typically with cerebral palsy or hypertonia, but it's criterion-referenced for children with Down Syndrome (DS) as well. This objective measure is more responsive to motor change over time with children with DS than the BSIDII. This measure helps more clearly define quality of change which can be very useful for insurance justifications.