Hypertonia

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Hypertonia is the abnormal increase in muscle tone as a result of upper motor neuron lesions. There are three following clinical types: spasticity, dystonia, and rigidity. Management of hypertonia is individualized and should be directed by the patient and/or family's goals of care as well as the underlying cause of the hypertonia. Treatment options include stretching, strengthening, positioning, oral medications, botulinum toxin injections, phenol injections, as well as surgical procedures. Without effective management, hypertonia can result in muscle imbalance, abnormal movement patterns, pain, joint contracture, joint deformity, and ultimately negatively impact a patient's function. This discussion serves as an overview of hypertonia, focusing on spasticity and dystonia, in the pediatric population by examining the causes and epidemiology, elucidating its symptoms, discussing available treatment and management options, and clarifying why this all matters.

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Introduction

Hypertonia is the abnormal increase in muscle tone as a result of upper motor neuron lesions. While hypertonia and spasticity are sometimes used synonymously, spasticity is actually just one of the three clinical types of hypertonia, in addition to rigidity and dystonia. In an effort to clarify potential confusion, the NIH Task Force on Childhood Motor Disorders established the following definitions:

Spasticity is defined as velocity dependent hypertonia.

Dystonia is defined as a movement disorder in which involuntary sustained or intermittent muscle contractions cause twisting and repetitive movements, abnormal postures, or both.

Rigidity is defined as hypertonia present at all rates of passive and active movement.[1]

While the treatment of spasticity, dystonia, and rigidity are all important, rigidity is rare in the pediatric population and will not be addressed in this article. The main difference between spasticity and dystonia is that spasticity is present at rest and occurs in response to passive movements, while dystonia is largely absent at rest and only becomes apparent during active movement. This discussion attempts to serve as an overview of hypertonia, focusing on spasticity and dystonia, in the pediatric population by examining the causes and epidemiology, elucidating its symptoms, discussing available treatment and management options.

Causes

Upper motor neuron (UMN) lesions disrupt the signal between the central and peripheral motor neurons, causing inappropriate/decreased regulation of downstream pathways. Any disease, disorder, condition, or trauma that causes an UMN lesion can cause hypertonia. While the exact lesions that cause spasticity are unknown and seem to vary, dystonia is often the result of cortical lesions of the thalamus or basal ganglia.[2–4] Conditions that commonly lead to hypertonia include cerebral palsy, traumatic brain or spinal injury, metabolic disorders, leukodystrophies, hydrocephalus, stroke, and multiple sclerosis.[4,5] Cerebral palsy is the most common cause of spasticity in children.[4,6,7]

The epidemiology of hypertonia is difficult to establish due to the large number of conditions that cause it, the variety of scales uses to measure it, and the fact that non-hypertonic comorbidities of certain conditions can present themselves as hypertonia when there is in fact little to no change in muscle tone.

Symptoms/Recognition

Spasticity

On clinical examination, spasticity presents with a number of positive clinical findings, such as hyperactive reflexes, extensor plantar responses, and clonus, as well as negative clinical symptoms, such as lack of agility, loss of

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selective motor control, fatigue, and poor coordination.[8,9] Patients also may present with retention of abnormal primitive reflexes such as the asymmetrical tonic neck reflex with sustained body postures from which the child cannot re-position him- or herself to exert voluntary control.[7,8]

Clinical assessment of spasticity should include attempted quantification. From a technical standpoint, the amount of spasticity present is affected by many extraneous factors, including ambient temperature, anxiety, time of day, level of pain, body position, and the amount of prior stretching, all confounding the interpretation of serial measurements.[8] One way to measure spasticity is to use an established scale or scoring system, of which many exist (Table 1).

Dystonia

Identifying dystonia in children can be complex, since it is classified as both a hypertonic condition as well as a hyperkinetic condition.[18] It can be difficult to differentiate dystonia from spasticity as well as other hyperkinetic movement disorders. Dystonia is characterized by involuntary, sustained muscle contractions involving opposing muscle groups (e.g., flexors and extensors at a joint) manifesting as repetitive movements, torsion, or abnormal postures.[13] Voluntary movements are slow and more variable than normal, due to co-contraction of inappropriate muscle groups, seen by an overflow of EMG activity.[14] Quantification of dystonia is also possible through the use of various scales (Table 1).

Treatment and Management

There is no one single treatment for hypertonia that is the gold standard; the type of treatment should be individualized depending on level of hypertonicity, the goals of the patient and his/her family as well as the family's ability to provide care, and the comorbidities of the underlying cause of the UMN lesion.[3,7,16]

TABLE 1. Scales used in the recognition and treatment of hypertonia[2,4,10–12,15]

Scale	Diagnoses	Ages	Type of test	Body region/function evaluated
Modified Ashworth Scale (MAS)	Hypertonia	All ages	Subjective passive ROM evaluation	Movement about a joint
Tardieu scale	Spasticity	All ages	Subjective passive ROM evaluation	Movement about a joint
Hypertonia assessment tool (HAT)	Spasticity Dystonia Tremor	4–19 years	Passive ROM evaluation	Upper and lower extremities
Burke–Fahn–Mardsen (BFM) scale	Primary dystonia	All ages	Subjective	Eyes Mouth/neck/trunk Bilateral upper and lower extremities
Barry-Albright dystonia scale (BAD)	Secondary dystonia	Not tested on children < 3 years old	Clinician observation	Eyes Mouth/neck/trunk Bilateral upper and lower extremities
Global dystonia scale (GDS)	Dystonia	All ages	Clinician Observation	Motor Function Oral/verbal function Self-care Attention/alertness Eye and periorbital region Face Mouth/neck/trunk Upper and lower limbs
Movement disorder-childhood rating scale (MD-CRS)	Movement disorder	<18 years old	Clinician observation parental subjective response	Motor function Oral/verbal function Self-care Attention/alertness Eye and periorbital region Face Mouth/neck/trunk Upper and lower limbs