Pediatric Tone Management

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KEYWORDS

• Hypertonia • Spasticity • Dystonia • Baclofen • Rhizotomy • Neurotomy
• Cerebral palsy • Pediatric

KEY POINTS

- The type of hypertonicity needs to be identified to determine optimal treatment.
- Pediatric tone management involves all individuals caring for that child, including the patient, family, therapists, and the medical team.
- There are multiple nonsurgical and surgical treatments for hypertonicity.

INTRODUCTION

Tone management is one of the primary roles of a pediatric physiatrist and is a rewarding but frequently challenging task. Hypertonicity frequently inhibits normal movement patterns in children with central nervous system (CNS) lesions. At times, hypertonicity can reinforce muscle group firing and be useful for a child's function, such as stabilizing the lower limbs during stand pivot transfers. Hypertonicity can manifest as spasticity, dystonia, or rigidity, and frequently a combination is present. The manifestations of hypertonicity, underlying etiologies, and guiding treatment principles are reviewed.

Spasticity is probably the most common and easily recognizable form of hypertonicity. Spasticity is defined as increased muscle tone where resistance to externally imposed movement increases with increased speed of stretch and varies with the direction of joint movement.[1] A child may experience difficulty with smooth movements because of spasticity. The muscle stretch reflex may be inadvertently triggered

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during activity, and the muscle "catch" may result in loss of postural stability. A commonly used scale to grade spasticity is the Modified Ashworth Scale using a range from 0 to 4, indicating no increased tone (0) to complete resistance to movement or rigidity (4).[2]

Dystonia is a more complicated form of tone. It is characterized as increased muscle tone due to abnormal involuntary co-contractions in muscle groups causing repeated abnormal posturing of the neck, torso, or limbs.[1] Dystonia is typically characterized as primary dystonia or secondary dystonia, which is typically due to an underlying cortical lesion in the thalamus or basal ganglia.[3] Functionally, dystonia or dystonic movements increase when a child attempts to perform a novel or difficult task. Usually when the child is relaxed, there is no increased tone and all limbs may be freely mobilized. When the dystonia is active, the affected body areas or limbs twist into varied postures even when the child is performing a task with the unaffected limbs. Severe dystonia with co-contractions can present at rest with joint rigidity. Dystonia is commonly measured using the Fahn-Marsden (or Burke-Fahn-Marsden) rating scale or the Barry-Albright Dystonia Scale. The Fahn-Marsden scale ranges from 0 to 4, from no dystonia, dystonia with a particular action, dystonia on many actions, to dystonia at rest. Each limb and the trunk, head, and neck are evaluated.[4] The Barry-Albright Dystonia scale also ranges from 0 to 4, but looks at the frequency of dystonia over 8 body regions.[5]

A mixture of spasticity and dystonia is frequently present in children with more severe CNS lesions and is important to consider when treating tone disorders. Spasticity, dystonia, or a combination can lead to limb rigidity, which complicates treatment options.

Many pediatric conditions lead to hypertonicity, including cerebral palsy (CP), acquired brain injury, metabolic disorders, leukodystrophies, hydrocephalus, or spinal cord injury. CP is the most common condition associated with hypertonicity.

CP "describes a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to nonprogressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of CP are often accompanied by disturbances of sensation, perception, cognition, communication and behavior, by epilepsy, and by secondary musculoskeletal problems."[6] Tone and disorders of movement vary greatly among the CP population but most have some difficulties with limb hypertonicity.[7,8] Cerebral vascular accidents, hypoxic ischemic events, hemorrhagic events related to extreme prematurity, traumatic brain injury, neonatal toxoplasmosis, other (syphilis, varicella-zoster, parvovirus B19), rubella, cytomegalovirus (CMV), and herpes infections (TORCH) infection, or isolated metabolic derangement injuries can lead to CP, highlighting the heterogeneity of this diagnosis. Neuronal migration abnormalities, such as schizencephaly, porencephaly, and polymicrogyria, may qualify under the CP diagnosis umbrella if no other genetic syndromes are associated.

When evaluating a child with newly noted hypertonicity, brain and spinal cord imaging should be considered to evaluate reversible etiologies. The American Academy of Neurology published a practice parameter guideline for "Diagnostic assessment of the child with cerebral palsy." This guideline summary provides levels of evidence associated with workup measures for a child with suspected CP.[9]

There are many causes of hypertonicity and a multitude of treatments to manage spasticity and dystonia. Treatment approaches should be individualized based on functional goals of the child and family, level of impairment, and/or ability to care for the child. The type, locality, and severity of hypertonicity need to be considered. Treatment plans should be created in collaboration with all individuals caring for the child, including the patient, family, therapists, and medical team.