The ABCs of Hypertonia Management – ITB, SDR, DBS

Cerebral Palsy is a disorder of muscle tone caused by a non-progressive brain insult with onset with onset from prenatal brain development through early childhood. Affecting approximately 3/1000 children, it is the most common disabling motor disorder beginning in childhood. With a wide range of etiologies, there is also significant variability in the motor presentation. Spasticity and dystonia are both common, and often there are overlapping abnormalities of tone. A common pattern is spasticity of the lower extremities, hypotonia of the trunk and axial musculature, with dystonia dominating in the upper extremities.

Spasticity is velocity dependent, typically unidirectional resistance to muscle stretch with associated upper motor neuron signs. It essentially involves disinhibition of GABA-ergic cells at the spinal cord.

Dystonia is hypertonia with stereotyped writhing patterned movements, often involving co-contraction of agonists and antagonists that may involve overflow to remote muscle groups. Biochemically, multiple neurotransmitters are involved through the basal ganglia and associated interconnections.

Treatment of spasticity is best managed by a comprehensive team approach that includes neurology, neurosurgery, orthopedics, rehabilitation specialists and support professionals. Initial tone management typically includes rehabilitation therapy, medications, neurotoxins and orthotics. Medications (many off label) may include baclofen, gabapentin, tizanidine and benzodiazepines. More severe patients may benefit from neurosurgical options focused on reducing tone at the spinal cord level. The chemistry of dystonia is more complex, thus the pharmacologic options are more varied and less consistently successful (table 1).

Pharmacologic options for dystonia

- Acetylcholine antagonist
- α Adrenergic agonist
- Dopamine precursor
- Dopamine agonist
- Dopamine antagonist
- Dopamine depletion
- Gaba-B agonist
- Gaba-A agonist
- Chemodenervation
- Trihexyphenidil
- Clonidine, Tizanidine
- Levodopa/carbidopa
- Ropinorole, Pramipexole
- Risperidone
- Tetrabenazine
- Baclofen, Gabapentin, Benzodiazepines
- Zolpidem
- Botulinum toxins A,B

Figure 1: Pharmacologic options available for the management of dystonia.
When pharmacologic management of tone is inadequate, neurosurgical options may be considered. Options are available for both generalized and regional tone management, and cross disciplines involving rehabilitative therapies, pharmacologic agents, neurosurgical options, and orthopedic surgery. Strategies for the management of hypertonia must take into account several factors, including:

- Etiology and expected natural history of the underlying pathology
- The nature and distribution of the hypertonia
- Presence of hypotonia and other abnormalities of tone or movement
- Associated co-morbidities (e.g. epilepsy, hydrocephalus, intellectual impairment)

Social factors, including needs and abilities of the family

Medication, primarily baclofen, can be delivered preferentially to the spinal cord via an implanted medication pump. While several systems are available, the most widely used is programmable allowing for dose titration. Placement of the catheter high or low in the spine helps target therapy depending on the degree of upper extremity involvement. Intrathecal baclofen may also reduce dystonia.

Selective dorsal rhizotomy, re-popularized in the 1980’s by Warnick Peacock involves the irreversible selective sectioning of sensory roots therefore decreasing feedback from spastic muscles to the lower to the spinal cord. Post-operative care includes extensive rehabilitation and retraining. Application of SDR is generally considered for ambulatory young patients with predominantly lower limb spasticity, and does not treat dystonia.

Deep brain stimulation (DBS) was introduced in the 1980s by Benabides and Pollack for the treatment of tremor. The US FDA granted HDE approval for the treatment of dystonia in patients 7 years of age and older in 2003. DBS involves the delivery of small electrical impulses directly to targeted areas of the brain. For dystonia, the primary target is the internal segment of the globus pallidus.

Once tone is adequately managed, orthopedic surgery may be needed to improve positioning and function. The majority of children with cerebral palsy and will live well into adulthood, thus early recognition and treatment of tone has lifelong implications for both patients and care providers. Goals of tone management include functional improvement, pain reduction and the prevention secondary complications including musculoskeletal contractures. Ultimately, success is marked by maximized personal independence and/or reduced caregiver burden.

As part of an overall management strategy, a multidisciplinary team can be invaluable. The importance of managing expectations through patient education is a key component to successful implementation of a treatment strategy for tone management (figure 2).
Figure 2: A treatment paradigm for the management of hypertonia.