

## Upper Limb Hypertonicity in Children with Cerebral Palsy: A review study on Medical and Rehabilitative Management

Rassafiani, Mehdi<sup>1</sup>, PhD.

*Pediatric Neurorehabilitation Research Center,  
University of Social Welfare and Rehabilitation Sciences, Tehran, Iran*

Akbar Fahimi, Nazila

*University of Social Welfare and Rehabilitation Sciences, Tehran, Iran*

Robab Sahaf, PhD.

*Iranian Research Center on Aging,  
University of Social Welfare and Rehabilitation Sciences, Tehran, Iran*

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Hypertonicity is the most common type of cerebral palsy consists of 85% of the affected children. It has a very complex nature making intervention and management very difficult. This article tries to make reader familiar with various types of intervention and introduce a new intervention process to help clinicians decide better. Literature was reviewed with two criteria including: identifying various interventions and their effects on upper limb hypertonicity and level of invasiveness of each intervention. This paper suggested a new way of looking at hypertonicity based on its two components (i.e., neural and biomechanical) and effectiveness of each intervention on these components. In the treatment and management of hypertonicity, clinicians are required to look at all aspects of hypertonicity and then based on the provided decision tree, decide which kind of treatment to be used for the child.

**Keywords:** hypertonicity, cerebral palsy, spasticity, intervention, decision making

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### Introduction

Upper limb (UL) Hypertonicity is a complex problem in children with cerebral palsy (CP) that needs to be fully understood in clinical decision making. Three main issues need to be taken into account in a successful model of intervention for upper limb hypertonicity, including components of hypertonicity, effectiveness of each intervention on the components, and level of invasiveness of the intervention. Based on these issues, a therapist can follow a stepwise management of hypertonicity.

Hypertonicity has two components, including neural (positive and negative symptoms) and biomechanical, along with their specific subcategories described in our previous article (1). All the components need to be considered when managing such a complex problem that is so important to expert occupational therapists (2-4). Furthermore, a therapist should look at the influences of various types of managements and decide which management is useful for each component. In other words, the management

strategies adopted for each of these components, differ based on the nature of the problem. For example, the biomechanical component of hypertonicity responds to treatment involving stretching, positioning, splinting and casting, but not to drugs, injections, and intrathecal administration of baclofen (5). Management of the negative symptoms of the neural component differ, and include neuro-developmental treatment, biomechanical approach and constraint-induced movement therapy. Table 1). summarizes the influence of different intervention methods involved in the management of UL hypertonicity on the neural and biomechanical components of hypertonicity The next important issue in the management of hypertonicity is the level of invasiveness of each intervention. Different methods of hypertonicity management can be classified on the basis of their degree of invasiveness see(Figure 1). Level of invasiveness refers to the level of harm that can be accompanied by each intervention. Using this parameter, hands on

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1- All correspondences to Dr. Mehdi Rassafiani, Email: <mrassafiani@yahoo.com>

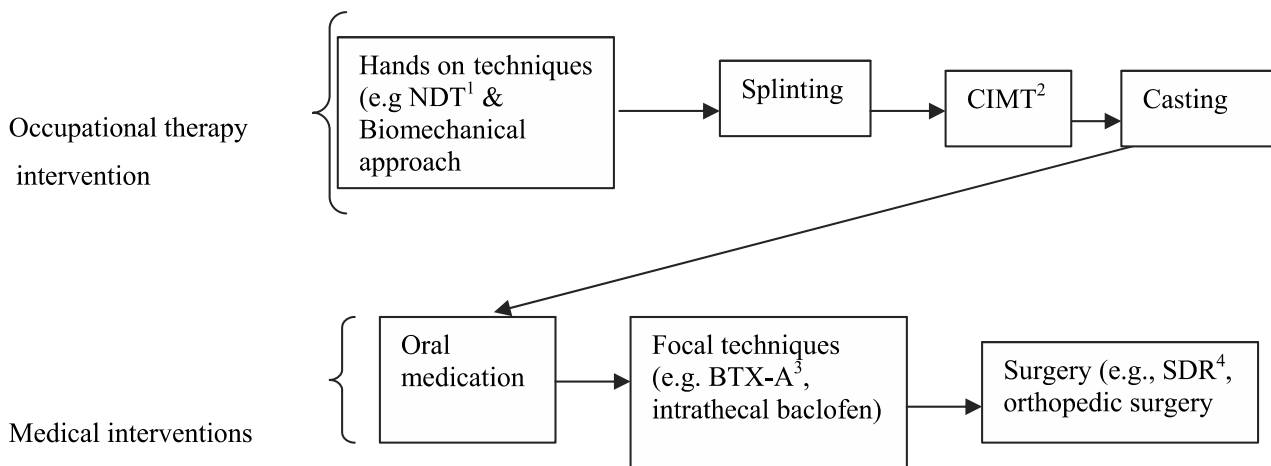
techniques can be seen as the least invasive at one end of a continuum and orthopedic surgery the most invasive at the other end (6). This classification helps occupational therapists identify an intervention method, but the most important question is how to choose the management method for a particular client.(Figure 2). presents an overview of the

hypertonicity management process based on the works of Barnes (2001) and Copley and Kuipers (1999) that helps therapists in their clinical decision-making by asking key questions (7). These intervention methods will be elaborated in the following section.

**Table 1.** The Intervention Methods and Their Influences on the Components of Hypertonicity

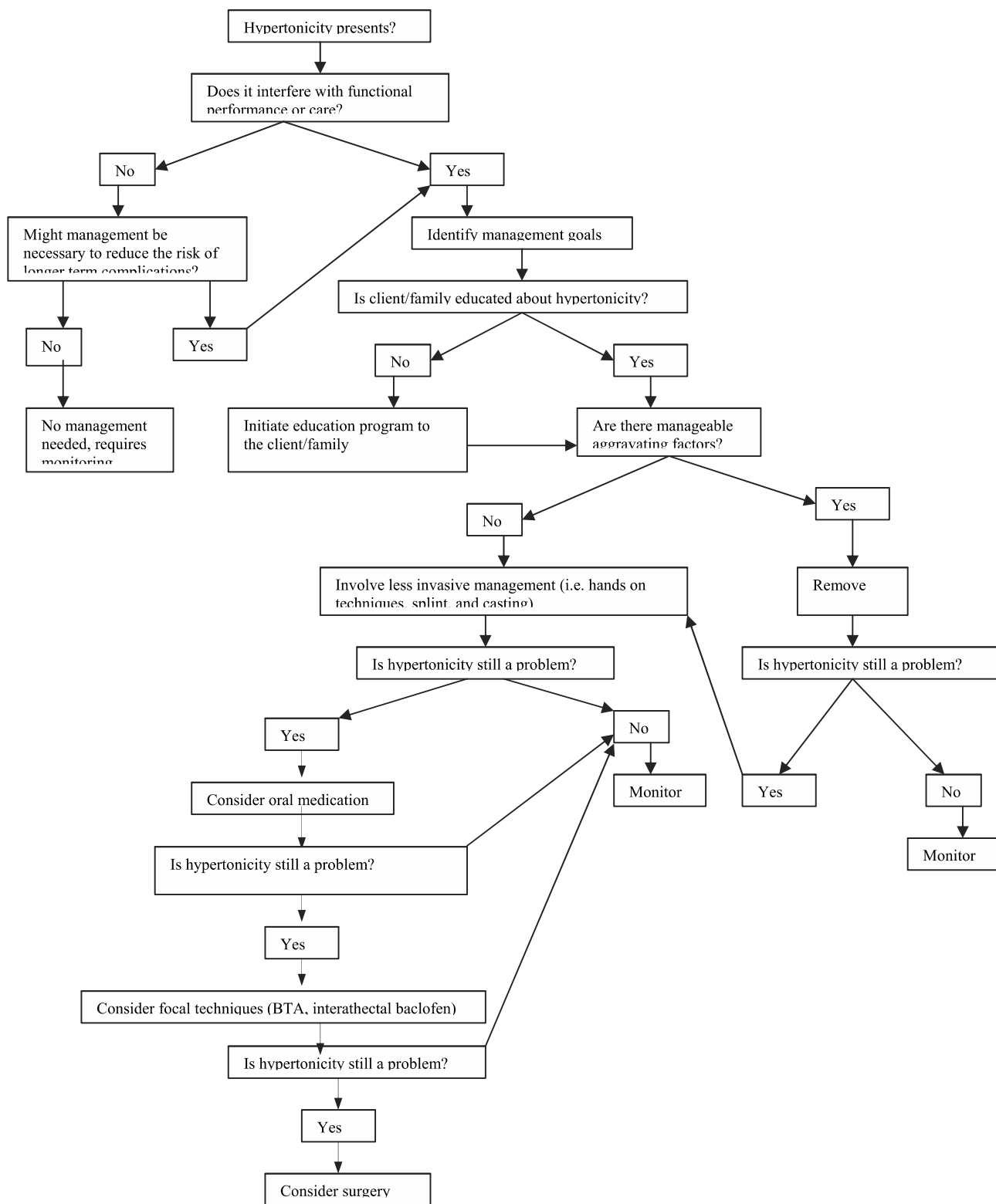
Components of Hypertonicity	Intervention Method								
	NDT <sup>1</sup>	BM <sup>2</sup>	SP <sup>3</sup>	CA <sup>4</sup>	CIM <sup>5</sup>	AD <sup>6</sup>	BTX-A <sup>7</sup>	ITB <sup>8</sup>	SU <sup>9</sup>
Neural: Positive symptoms	√	√	√	√	√	√	√	√	√
Neural: Negative symptoms	√	√			√				
Biomechanical components	√	√	√	√					√

<sup>1</sup>Neuro-developmental treatment; <sup>2</sup> Biomechanical approach; <sup>3</sup>Splinting; <sup>4</sup>Casting; <sup>5</sup>Constraint-induced movement therapy; <sup>6</sup> Antispastic Drug; <sup>7</sup> Botulinum Toxin-A; <sup>8</sup> Intrathecal Baclofen; <sup>9</sup> Surgery.



**Figure 1.** Various managements of hypertonicity from the least invasive (hands on techniques) to the most invasive (surgery)

1- Neuro-Developmental Treatment; 2- Constraint-Induced Movement Therapy;  
3- Botulinum Toxin-A; 4- Selective Dorsal Rhizotomy



**Figure 2.** Flow chart outlining the approach to the overall management of hypertonicity (adapted from Barnes, 2001, p.8; Copley & Kuipers, 1999)

## **Hands on Techniques: Neuro-developmental Treatment**

Various frames of reference and approaches to the treatment of neurobiological dysfunction have been developed since the 1940s. Neurodevelopmental treatment (NDT) is one of the most commonly used frames of reference in the management of UL tonal abnormalities (8). NDT was developed by Berta and Karel Bobath in the 1940s. Originally, it was developed for the treatment of children with neurological impairments, especially those with CP (9). However, NDT is now utilized for clients (children and adults) with movement disabilities such as neuromuscular disorders, and developmental disabilities (10).

NDT was previously based on two important principles: (1) the use of reflex inhibiting postures (RIP) aimed at decreasing muscle tone by the use of postures to oppose the primitive reflex patterns typically assumed by the child and, (2) the facilitation of normal motor development through righting and equilibrium reactions (11). The aim of facilitation was to help clients achieve motor developmental milestones. Current theoretical foundations of NDT include an in-depth understanding of normal development, components of movement, and atypical development (11). These theoretical foundations help therapists to critically evaluate clients with CP, as well as to develop appropriate treatment planning.

### *Normal development:*

NDT adopts a developmental frame of reference and while it was initially assumed that normal motor development passed through a cephalo-to-caudal, proximal-to-distal, and gross-to-fine sequence, this has been replaced by the interactional idea, in which development of a motor milestone depends not only on improving the control of the distal part of the body, but also at the same time on the proximal parts (11). For example, developing sitting control is dependent on a certain degree of pelvic and lower limb control. In other words, the head and trunk, and lower limbs interact in achieving control, alignment, and posture. Therefore, development occurs through an interaction between proximal and distal control (10).

Two other important concepts of normal development in NDT consist of mechanisms of 'feedback' and 'feed-forward'. Feedback is used to refine movement during a task. For example, while a person is writing, s/he may use proprioceptive, touch and visual information to correct her/his movements (11). Feedback can be either external (knowledge of

results) or internal (knowledge of performance) (10). NDT puts an emphasis on feedback from three primary sensory systems- vestibular, visual, and somatosensory- during the execution of movements (10, 11). Feed-forward is the anticipation of movement and postural control that is very important in functional activities (11). Feed-forward happens, for example, when a person decides to pick up a mug. If the person anticipates that an empty mug is full, s/he will apply more effort to picking it up than if the mug is thought to be empty.

The third concept of normal development in NDT involves postural alignment, postural control, and the base of support. Postural alignment is the ability to maintain a center of gravity over the base of support. The base of support is that part of the body that makes contact with the support surface (11). For example, in standing, the plantar surfaces of the feet form the base of support. Postural control is the ability to assume and keep postures during static and dynamic movements, while postural alignment is a prerequisite of postural control.

### *Components of Normal Motor Development:*

Normal motor development has three components: (1) interaction between stability and mobility, (2) postural tone, and (3) ability to dissociate movements. Stability involves retaining a posture against opposing forces (11). The point of stability is the body part in contact with the support surface. For example, in the quadruped position, all four limbs provide stability to the person. Mobility refers to smooth, controlled, coordinated movement based on a point of stability. The point of stability provides a base of support to the person to achieve a weight shift in any direction. This enables the person to move and manipulate the environment (11). For example, when in a quadruped position, a child can shift weight onto his/her legs and left arm, in order to free the right arm to reach out for a toy.

NDT emphasizes postural rather than muscle tone. Postural tone is defined as the distribution of tone throughout the body rather than in specific muscles. Normal postural tone is an amount of tone that is low enough to allow movement against gravity (mobility) and at the same time is high enough to maintain a stable posture against gravity (stability). Another component of normal development is the ability to dissociate movement, i.e. the ability to differentiate movements between the various parts of the body. This ability occurs within the first year of life and is a demonstration of CNS maturation. It is illustrated by separate movements between parts of the body and

within a given part. In a normal infant, a variety of movements can be observed when interacting with objects in the environment. This variety of motor development is the basis for the performance of functional skills in which the person pays attention to the goal of the movement rather than on the performance of the movements (11).

#### *Atypical Motor Development:*

There are two assumptions underlying atypical motor development, first, the disruption in feedback and feed-forward mechanisms, and second, abnormal postural control. Traditionally, NDT was modeled on the hierarchical structure of the CNS in which there are four levels of postural reflexes and reactions (i.e., spinal, brainstem, midbrain, and cortical level) (10). It was believed that abnormal movements were caused by the persistence of primitive reflexes or compensatory attempts to gain antigravity control (11) as well as sensory feedback (10). However, this model has been changed to a 'distributed control model'. Based on the distributed control model, the CNS is viewed as a system capable of initiating, anticipating and controlling movements (10). The CNS is not a passive system, but an active one using feedback and feed-forward mechanisms to control posture and movement. It is currently believed that abnormal movements are the result of a disruption in the feedback and feed-forward mechanisms. This impairs the acquisition of postural control and changes the experience of learning movement skills (10, 11).

Another assumption for abnormal movements is related to problems in postural tone (11). For example, increased muscle tone can limit movements causing a reduction in the degree of freedom of joints. This limitation leads to shortening of muscle fibers and contractures. Abnormal postural tone also interferes with movements causing total pattern of movements (flexor or extensor) (11). For example, when the person wants to reach out to pick up something, instead of coordinated and well-controlled movement in the upper limbs, a whole flexor pattern can be seen that is sometimes followed by abnormal head and trunk movement. This is a kind of compensatory pattern of movement resulting from abnormal postural tone which the person employs to achieve some degree of independence in functional activities.

#### *Intervention:*

Intervention within a NDT frame of reference occurs through handling, preparation, and facilitation (11).

Handling is graded sensory input provided by the therapist's body, mostly hands, at key points of control. It is used for preparation and facilitation of active postural control and movement patterns. Preparatory activities aim to promote mobility in those areas with limitations in passive range of movements and facilitate alignment of the body before active movement. Based on improved body alignment, muscle contraction around joints enables stability and mobility to be achieved. Consequently, movement and postural control are developed (11). In other words, through handling, preparation and facilitation, therapists utilize selective tactile and proprioceptive input to produce alignment, gain elongation of muscles, or facilitate normal muscle contraction and movement (12). Therapists usually employ compression, traction, deep pressure, weight bearing, and weight shifting in the intervention process and handling to facilitate motor control (11). Two important considerations in the management of clients involve active participation and gradual withdrawal of direct input by the therapist (10, 11). NDT sessions should be designed to include functional activities that are important to the person and encourage participation. These functional activities, which use goal-directed feed-forward mechanisms, should include activities of daily living (e.g., dressing, feeding), and play (10).

#### *Effectiveness of Neurodevelopmental Therapy:*

Only a few studies have been conducted to investigate the effectiveness of intensive and regular NDT, with or without other types of therapy on upper limb hypertonicity of children with CP. Kluzik, Fetters, and Coryell (1990) studied the influence of NDT on five children with hypertonicity (13). The results indicated that NDT improved the kinematic properties of reaching. The smoothness of reaching improved and reaching movements became significantly faster following the intervention. In a similar study, Fetters and Kluzik (1996) investigated the effectiveness of NDT on the reaching behavior of eight children with CP (14). The results supported those of Kluzik et al. (1990). However, small sample sizes limit the confidence with which these findings can be generalized. Moreover, these studies did not show the influence of NDT on functional outcomes.

Law et al. (1991) examined the influence of intensive (two times therapy per week and 30 minutes daily home program) and regular (once-weekly therapy and 15 minutes daily home program)

NDT, and casting on hand function, range of movement, and quality of movement of 72 children with hypertonic CP aged between 18 months and 8 years (15). The results demonstrated no differences between the two interventions; however, casting improved the results of the NDT. In another study, Law et al. (1997) investigated the effectiveness of NDT plus casting and occupational therapy on the hand function of 50 children with CP aged between 18 months and 4 years (16). The results demonstrated that both programs led to a significant improvement in hand function, quality of upper limb movements, and parents' perception of hand function, but no significant differences were obtained between the two interventions. These findings suggest no beneficial effects were achieved by intensive NDT alone for the children in the study. The effectiveness of NDT on upper limb hypertonicity requires further investigation due to limitations in the current literature (17). In addition, the application of NDT has changed over the last two decades; therefore its effectiveness needs to be further examined. It is also necessary to compare different intensities of NDT, as well as the results of NDT interventions when used in conjunction with other types of therapy.

### **Biomechanical Frame of Reference**

The biomechanical frame of reference addresses the implications of physical and physiological theories on motor development (18). It applies the principles of kinetics and kinematics to the movements of the human body (19). This frame of reference aims to improve range of motion, muscle strength, and endurance, and reduce deformity or decrease the effects of deformity (19-21) in people whose capacities for functional motion are reduced.

The biomechanical frame of reference is based on five assumptions (18). First, a biomechanical approach provides external supports to substitute for inadequate or abnormal postural reactions. In this way it contributes to independence. Second, it assumes that motor patterns develop from sensory stimulation. In infancy, movements are mostly reflexive and result from tactile, proprioceptive, or vestibular stimuli. These reactions, in turn, provide additional sensory input helping children to develop motor control. Third, automatic motor responses which maintain posture develop in a predictable way. Righting and equilibrium reactions help children to move to a position, and maintain that position. The biomechanical frame of reference

concentrates on function within a relatively static position (i.e., supine, prone, side-lying, sitting, and standing) rather than dynamic situations. Fourth, the development of normal postural reactions is influenced by dysfunction or abnormalities of muscle tone, bone, or the central nervous system. Lastly, the level of postural dysfunction and external factors that may affect performance must be determined.

The biomechanical frame of reference is mostly employed to treat people with orthopedic disabilities (18). However, its basic concepts are utilized in fabricating splints and casting to prevent or reduce contracture and deformity in the limbs of clients with hypertonicity. Intervention in the biomechanical frame of reference happens through activities that provide passive stretching of muscle, passive joint ranging (6, 22), positioning, head and trunk control, as well as functional skills that promote head and arm movements, mobility, feeding, and toileting (18).

### **Splinting**

Splinting (for the upper limb) involves the use of any medical device applied to or around the upper limb to address physical problems (23). Splinting aims to maintain muscular balance between hypertonic muscles and their antagonists, to prevent muscle and joint contracture and deformity, to reduce hypertonicity and assist function by positioning the upper limb, to reduce edema, manage skin breakdown, and to maintain upper limb appearance (6, 24).

Mechanisms of splinting can be explained through the biomechanical, neurophysiological, and cognitive motor learning frames of reference (6). Based on the biomechanical frame of reference, splints provide passive stretch, joint stability, maintain joint alignments, and reduce pain. According to the neurophysiological frame of reference, splints can reduce hypertonicity through applying pressure on the insertion of muscle tendons and constant stretching over the hypertonic muscles, and providing neutral warmth to the limb that is splinted. In addition, splints assist people with hypertonicity to improve positioning and achieve more normal movement patterns leading to more effective function. There are various classifications for splints based on function and the major joints included (24). However, the most important splints employed for hypertonicity include resting hand splints, functional hand splints, elbow splints, static splints, dynamic splints, and semi-dynamic splints (6).

### Constraint-Induced Movement Therapy

Constraint induced movement therapy (CIMT) or forced use training has been used as a method of treatment for clients with stroke-induced hemiplegia (25-27), and recently for children with CP (28-30) to improve upper limb function. Charles and Gordon (2005) reviewed literature and identified a moderate success in motor performance following CIMT (31). This technique employs motor restriction of the less-affected or non-affected arm, as well as intensive training of the more-affected arm (25). Results to date suggest improvement in upper limb function in the areas of reaching, grasping, releasing, and weight bearing. It also reportedly assisted children to develop motor abilities needed for play and daily activities. However, the influence of CIMT requires more investigation to understand the characteristics of children who benefit from the intervention. Moreover, it is essential to study possible adverse side effects of this intervention (i.e., the client's emotional distress) (32).

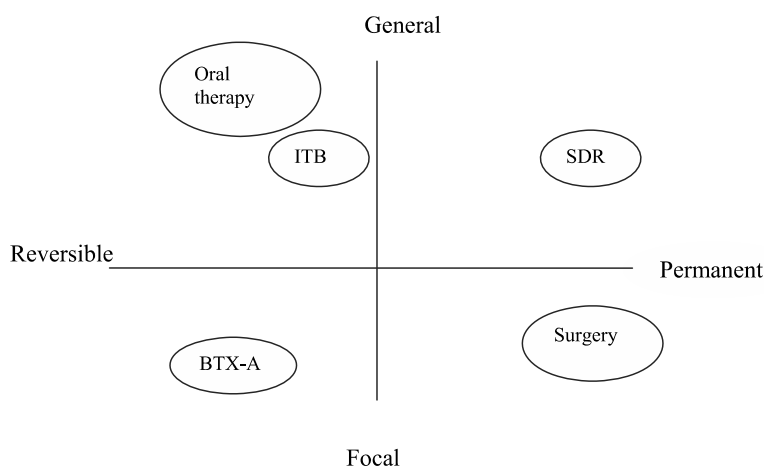
### Casting

Casting is another adjunct technique employed to reduce hypertonicity and contracture. Casting has been shown to improve range of movement and decrease exaggerated muscle tone resulting from hypertonicity (33). There are two mechanisms underlying the effectiveness of casting: mechanical and physiological (15). The biomechanical mechanism of casting helps to promote muscle

lengthening and joint range of movement by reducing soft-tissue contractures resulting from long-term hypertonicity (34). The physiological mechanism of casting is through provision of neutral warmth and cutaneous pressure, and therefore reduction of hypertonicity (15, 35). Suitable clients for casting can be divided into two groups (36). The first group is characterized by high physical disability, little functional use of the limb, and intellectual impairment. Casting aims, in this group, to reduce hypertonicity and contracture in order to make hygiene management and positioning for the caregivers easier. The second group comprises clients who have more active, functional use of the upper limb and are more cognitively aware. Casting for these clients aims to improve functional use of the limb in activities of daily living.

### Medical Management

Medical management of hypertonicity is complex, multi-professional (37) and includes pharmaceutical, neurosurgical, and orthopedic surgery. Choosing among these strategies to control hypertonicity depends on a number of factors including: age, severity of spasticity, cognitive abilities, and preference of family and/or doctor. The results of these methods can be permanent such as orthopedic surgery or reversible such as taking Valium. These methods can be either general, such as selective dorsal rhizotomy (SDR) or focal, such as Botulinum toxin - A injection (see Figure 3) (38).



**Figure 3.** Roles of different methods of the management of hypertonicity available for CP (adopted from Graham et al., 2000, p.71)

Note. ITB: Intrathecal Baclofen; SDR: Selective Dorsal Rhizotomy; BTX-A: Botulinum Toxin- A.

### Pharmacotherapy

Pharmacotherapy is a temporary management strategy (38) regarded as additional rather than

substituent for physical management (39) of hypertonicity in children with CP. It is easy to use with a short term effect which can be given as focal

treatment such as botulinum toxin -A (BTX-A) injection or generalized such as intrathecal baclofen. Indications for pharmacological treatment of hypertonicity include: (1) increasing tone despite physical stretching or casting; (2) pain due to hypertonicity; (3) prevention and treatment of contracture formation; (4) prevention of deformities; (5) management of skin hygiene; (6) cosmetic effect; and (7) decreasing caregiver burden for performing caregiver tasks (37). Medications may also help to address hypertonicity in various methods and include oral drugs, chemical neurolysis, intrathecal baclofen, and BTX-A (37, 40).

Oral anti-spastic agents are generally given in the presence of widespread hypertonicity rather than a local problem (37) and consideration of a number of issues is required. First, clear goals need to be determined and communicated to clients in order to ensure appropriate expectations (41). These goals should be part of the rehabilitation process (37). Second, the drug's side effects should be explained to the client. Third, anti-spastic drugs are usually introduced at low doses and incrementally increased to a point where it is clinically effective. Fourth, most anti-spastic drugs can be given in combination with each other to improve the clinical effects and decrease side effects. Finally, it is important to measure the outcome of drugs on the client's hypertonicity to see whether the goals of treatment have been achieved (37). The most commonly used drugs for the reduction of hypertonicity include baclofen (Lioresal), diazepam (Valium), tizanidine, and dantrolene sodium, (40, 42).

#### *Chemical neurolysis:*

Chemical neurolysis is another method used to reduce muscle hypertonicity. This method relieves hypertonicity in most cases without significantly affecting voluntary movement (43). In this method, phenol or alcohol is employed in three ways to provide chemical neurolysis including: peripheral nerve blocks, motor point (intramuscular) injections and the intrathecal administration (44). These procedures are preferred to oral anti-spastic agents, which often cause general adverse effects influencing both normal and hypertonic muscles (44). Phenol or alcohol injection is indicated in UL hypertonicity to facilitate activities of daily living, and improve hand hygiene. However, it may result in vascular damage and loss of skin sensation. Therefore, BTX-A is preferred to phenol and alcohol for the management of UL hypertonicity (44).

The intramuscular injection of BTX-A is becoming a popular procedure to reduce hypertonicity in children with CP, and others. It causes a local temporary muscle paralysis (45, 46) by inhibiting presynaptic release of acetylcholine at the site of the neuromuscular junction (47) leading to decreased hypertonicity with minimal side effects (38). BTX-A has also been reported to have positive effects on cosmetic appearance (e.g., it helps correct the "thumb in palm" posture) (48), quality of functional movement of upper limb and functional capability in children with CP (46, 49). BTX-A is also helpful in surgical decision making for children with hypertonic CP (50). When BTX-A is injected prior to surgical intervention, it results in a condition similar to tendon or muscle lengthening and tendon transfer. This provides suitable information to the surgeon to make decisions more efficiently.

BTX-A is indicated when a child has moderate hypertonicity, no fixed contractures, fairly good sensory awareness, and the ability to initiate movement in the hand, as well as adequate grip strength prior to injection (38, 46). However, these characteristics require more in-depth research to understand the criteria for indication of BTX-A injection and a generally accepted treatment protocol (38). Moreover, because the effects of BTX-A only last four months (47), it is necessary to examine the long term effects as well as the effects of several injections over a long period of time. Although, BTX-A is an adjunct to therapy (51), the influence of BTX-A plus occupational therapy, physical therapy, splinting, casting etc, requires further investigation to better determine any interactions and the impact on various aspects of disability such as strength, passive range of motion, abnormal patterns, functional outcome, and activities of daily living.

As previously mentioned, Baclofen can be delivered orally to reduce hypertonicity. Additionally, it can be administered intrathecally (Intrathecal Baclofen Therapy or ITB) (T4-T6) to reduce hypertonicity in upper limbs (37). Baclofen inhibits both monosynaptic and polysynaptic reflexes causing a reduction in spasticity (37, 52). It is used in clients with widespread hypertonicity, and those in which alternative methods of management are ineffective and inadequate, or cause unacceptable side effects (52).

Almedia et al. (1997) used a single subject experimental design to examine the effect of ITB on a child with spastic diplegia over two years (53). They demonstrated that ITB decreased spasticity, but had no effect on range of movement. Upper limb



movement speed, as well as dressing and the ability to transfer improved. Since these results are based on just a single case, more investigation is required. Moreover, in the long term, the influence of maturation needs to be considered. Albright, Barron, Fasick et al. (1993) examined the effects of ITB on upper limb hypertonicity of cerebral origin of 37 people aged 5 to 27 (54). They showed that ITB reduced hypertonicity, and promoted upper limb function, particularly it greatly improved the ability to reach upward and supinate. It also improved activities of daily living (ADL). Methodological limitations such as the absence of a control group limit the confidence with which these results can be interpreted. In addition, there were no standard assessments to assess ADL and functional changes.

### Surgical Management of Hypertonicity

When hypertonicity is not controllable by drugs, BTX-A, ITB, or chemical neurolysis, it may benefit from neuro-ablative procedures, or orthopedic surgery (55). Neuro-ablative procedures involve selective lesions that can be performed at the level of peripheral nerves (peripheral neurotomy), or spinal roots (dorsal rhizotomy) (55). Peripheral neurotomies (PN) are a technique indicated when hypertonicity is localized to muscle groups supplied by a single or a few peripheral nerves that are easily accessible (55). In the upper limb, PN may be performed in the musculocutaneous nerve for hypertonic elbow flexion; in the median and ulnar nerve for hypertonic wrist and finger flexors; or in

the brachial plexus branches for treating hypertonic shoulder. PN helps to reduce hypertonicity and deformity as well as improve motor function by rebalancing the actions of agonist and antagonist muscles. Selective dorsal rhizotomy is another technique used to cut selective dorsal roots in the spinal cord to decrease hypertonicity (55). Selective dorsal rhizotomy is mostly employed to reduce hypertonicity in lower limbs rather than in upper limbs (56).

Orthopedic surgery aims to reduce pain, prevent or correct deformity, and improve function, cosmetic and hygiene (57). Common orthopedic procedures include tendon release, muscle lengthening, tendon transfer and osteotomies (55, 58). In the upper limb, flexor digitorum lengthening is a common procedure used for a hemiplegic hand to obtain a more functional position. Tendon transfer is employed to normalize articular orientation in case of muscular imbalance (55). Osteotomies are another type of surgery that aim to correct bone deformity resulting from growth distortion used mostly for lower limbs deformities (55).

### Summary

This review provided a new model of practice in children with upper limb hypertonicity based on the components of hypertonicity and the level of invasiveness of the intervention. These components were used to help structure an outlook of intervention strategies currently used in the management of hypertonicity.

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