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CARE PATHWAYS

CENTRAL HYPOTONIA

Authors (AACPDM Central Hypotonia Care Pathway Team): G. Paleg (lead), R. Livingstone, E. Rodby-Bousquet, M. Story, and N.L. Maitre

DEFINITIONS

Hypotonia can be defined as abnormally low muscle tone, or reduced resistance to passive, relatively rapid movement. The imprecision of the definition reflects the lack of psychometric properties and reliability of assessments for hypotonia, therefore only clinical definitions currently in use by neurology specialists will be used in this pathway. Other terms for hypotonia include, but are not limited to, central hypotonia, floppy baby syndrome, benign congenital hypotonia, and neonatal hypotonia.

Hypotonia may originate from disturbances in the physiology of central or peripheral nervous systems or of the end organs themselves (muscles and muscle groups). The current pathway will only address children whose hypotonia is centrally-mediated and will exclude those whose hypotonia can be attributed conclusively to peripheral causes. Spinal Muscular Atrophy (SMA) is not included within the definition of centrally mediated hypotonia. Centrally-mediated hypotonia will be further referred to in this document as "central hypotonia" in the interest of simplicity. Specific etiologies of central hypotonia include brain insults and malformations, as well as genetic, metabolic, traumatic, anatomical, or idiopathic causes of central neural dysfunction.

Central hypotonia may be generalized and affect the limbs, trunk and neck or may be localized such that specific areas of the body are predominantly hypotonic with others having normal or hypertonic characteristics. Hypotonia is often seen in combination with muscle weakness. In the case of perinatal insults to white matter tracts, such as in encephalopathy of prematurity and neonatal encephalopathy, central hypotonia can evolve over the course of the first few years of life and progress to hypertonia. In addition, central hypotonia can co-exist with abnormalities of movement (such as ataxia or dyskinesia) or sensation (dysethesias, paresthesias).

WHY IS THERAPEUTIC ASSESSMENT AND INTERVENTION IMPORTANT FOR CHILDREN (AGE 0-6 YEARS) WITH CENTRAL HYPOTONIA?

Infants and young children with diagnoses of Down syndrome (DS), Cerebral Palsy (CP), and/or developmental delay (DD) often present with low muscle tone that can influence their gross motor development. Other children presenting to therapists may have no established diagnoses.

- Central hypotonia can impede motor function through decreased joint stability, joint hypermobility, weakness, and/or decreased activity and endurance.
- Impaired motor function can be associated with reduced developmental experiences in turn altering typical progression of gross and fine motor abilities.
- Central hypotonia can interfere with ability to attain positions and acquisition of developmental milestones.
- Hypotonic postures can interfere with functional activities such as reaching, sitting, standing and crawling/walking, which can lead to participation restrictions.
- Central hypotonia, in combination with muscle weakness, can interfere with sleep by limiting ability to change position during rest: this can contribute to discomfort and poor sleep quality.
- Central hypotonia can result in reflux and/or constipation due to abnormalities in coordination of voluntary and involuntary muscle function.
- Hypotonic postures and low muscle activity can create challenges for care-giving and participation in daily life activities.
- Children with central hypotonia can have drooling and feeding problems (e.g. chewing or swallowing).

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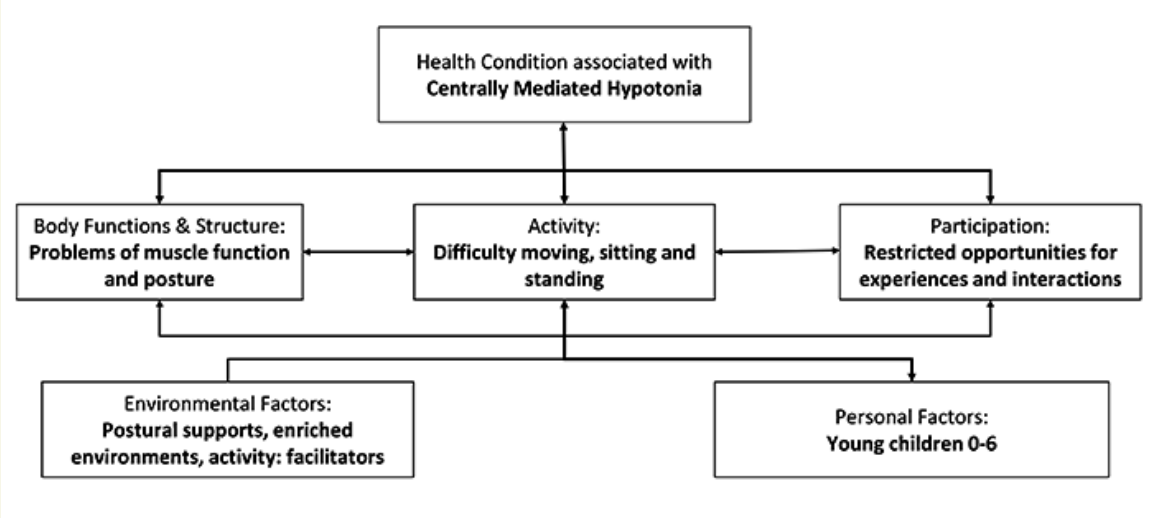

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Target Population: Children from birth to six years-of-age with central hypotonia and gross motor delays that limit activity and participation.

Target Clinical Providers: Therapists/Early Interventionists caring for children from birth to six years-of-age with central hypotonia and their families.

ASSESSMENT

If the child has not already been seen for a diagnostic work-up, referral to a neurologist, geneticist and/or developmental medical specialist is always recommended. During the therapy assessment, determine the impact of the hypotonia on function, activity limitations and participation restrictions, pain/comfort (including sleep), care-giving and whether management is required. Assess whether the clinical presentation is consistent with infants at "high risk of CP" (see Early Detection Guidelines, JAMA Pediatrics Novak et al. 2017).

Therapy Assessment: Children with hypotonia may have delays in motor development. It is recommended that therapists use valid and reliable measures of motor abilities. As there are no established evidence-based approaches to measure hypotonia, and as the focus of therapy should be on improved functioning, we recommend the following: use motor function assessments with good psychometric properties for infants at high risk for motor delays and neuromotor problems such as: the Hammersmith Infant Neurological Examination (HINE, 3 to 24 months), the Test of Infant Motor Performance (TIMP, term to 3 months), Peabody Developmental Motor Scales (PDMS), Development Assessment of Young Children (DAYC-2) Motor Scale, the Alberta Infant Motor Scale (AIMS, 0-18 months), Brigance III, etc. (See Early Detection Guidelines for CP, JAMA Pediatrics Novak et al., 2017). A broader perspective on promoting child development is offered by the 'F words in childhood disability' (Rosenbaum & Gorter, 2012)

Goal Setting: Use valid and reliable outcome measures outcomes such as Canadian Occupational Performance Measure (COPM), Goal Attainment Scaling (GAS) (see Section III for further details).

MANAGEMENT

Most of the interventions in the Central Hypotonia Care Pathway have low or very low levels of evidence (based on GRADE ratings).

Developmental Strategies: strategies used by physiotherapists (PTs), occupational therapists (OTs) and early interventionists are considered cornerstones in the management of central hypotonia. General principles include:

1. ensure therapy is goal-directed and promotes function and participation in daily activities/ routines.
2. ensure all motor interventions are child active.
3. activities should be child-initiated and child-directed.
4. activities should be caregiver delivered when possible.
5. optimize seating and upright positioning with good stability/support as early as possible (providing opportunities for reach/grasp and manipulation to learn through play).
6. avoid extreme positions (e.g. frog-legged (hip/knee flexion with abduction)) and strive for symmetry.
7. consider orthoses and splints to increase foot stability in stance and weight bearing.
8. encourage early mobility using various typical, adapted and specialized equipment.
9. coach parents to integrate therapeutic interventions for hypotonia into daily life and routines.
10. avoid passive interventions in which the therapist performs the work for the child and/or the child is not moving actively (reduce hands-on time and overt assistance, allow for infant-initiated activities).

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11.avoid interventions for which there is no evidence and/or a risk of negative outcomes (see section II for further details).
12.ensure screening for other health concerns and comorbidities including; with vision, hearing, feeding, reflux, and communication.

THERAPEUTIC RECOMMENDATIONS

(for details on each intervention including potential risks, see the evidence summary in Section II):

1. Tummy Time activities (during supervised play, when a child is awake), for multiple short sessions per day, may promote motor development in young children with central hypotonia.
2. Active motor abilities should be promoted in sitting, standing and for mobility.
3. Infant massage may be used to promote mother-infant bonding and sleep.
4. Treadmill training may be used from 10 months onward, to promote early onset of stepping, walking and improve gait characteristics in children with central hypotonia.
5. Orthotics may be used to support foot alignment and improve gait characteristics for ambulatory children with central hypotonia; in pre-ambulatory children, expert opinion recommends trial and/or use of orthoses when ankle instability prevents age appropriate exploration.
6. Adaptive equipment may increase activity and participation: e.g. adaptive seating; compression garments, walker/gait trainer; stander; and power mobility devices.
7. Postural management programs facilitate age appropriate activity and participation in natural routines (i.e. activities in lying, supported sitting, standing). Postural management programs should reduce time spent in asymmetrical lying postures and frog-legged positions.
8. Hip surveillance to monitor hip displacement can enable referral for early intervention to prevent hip subluxation and dislocation, which is known to occur in children with central hypotonia (see AACPDM Hip Surveillance Care Pathway for information on surveillance in children with cerebral palsy).

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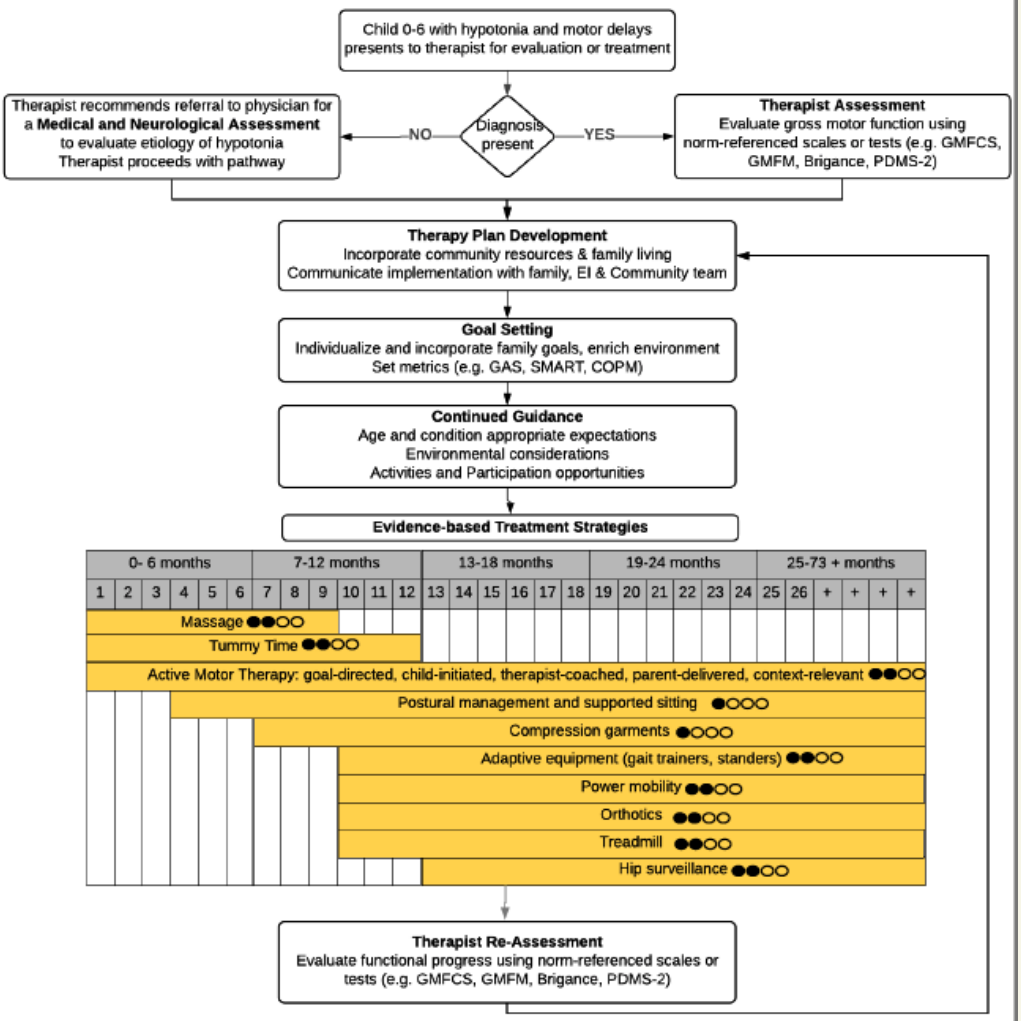
Hypotonia Care Pathway Algorithm



Legend:
Yellow color refers to nature of recommendations in GRADE- conditional for all
Evidence levels:
Very Low ●○○○ Low ●●○○ Moderate ●●●○ High ●●●●

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