DYSTONIA

Bottom Line ‘Evidence-Informed’ Recommendations for the Management of Dystonia in Individuals with Cerebral Palsy


DEFINITIONS
Dystonia is a movement disorder in which involuntary sustained or intermittent muscle contractions cause twisting and repetitive movements, abnormal postures, or both. Dystonia in cerebral palsy (CP) presents as hypertonia, involuntary postures and movements, or a combination. Dystonia occurs in dyskinetic CP but also is commonly present in spastic CP.

WHY IS DYSTONIA IN CEREBRAL PALSY IMPORTANT?

- Dystonia can impede motor function through involuntary muscle contractions, limitations in muscle relaxation, and overflow, which is the association of involuntary movement with intended movement which spreads to surrounding or distant muscles
- Dystonia can interfere with positioning for sitting and lying
- Dystonic postures and movement can be painful
- Dystonia can interfere with sleep
- Dystonia can result in high energy expenditure and malnutrition
- Dystonic postures/hypertonia can create challenges with care-giving

Target Population: Individuals with dystonia in CP where the dystonia interferes with function, positioning or causes pain and disrupted sleep.

Target Clinical Providers: Physicians/Nurses/Therapists caring for individuals with dystonia in CP.

ASSESSMENT
Dystonia is a frequently overlooked element of the neurological presentation of CP. Therefore, it is recommended that a ‘dystonia’ assessment be routinely included in your neurological examination (assessing for fluctuating hypertonia, and using tactile stimulation or voluntary movement to trigger dystonia). More information can be found in Section 3 of this pathway in the HAT tool link. On your examination, determine if the dystonia is generalized or focal and assess the severity (can use a standardized scale such as the Barry Albright Dystonia Scale outlined in Section 3). Assess the impact of the dystonia on function, pain/comfort (including sleep), and care-giving and whether management is required.

If dystonia is present, assess whether the neurologic presentation is consistent with CP (risk factors, brain imaging, and family history) or if additional work-up is required. An important masquerader of CP-related dystonia is Dopamine Responsive Dystonia. Consider the need for a trial of levodopa and/or a referral to a neurologist/geneticist for additional diagnostic work-up.

MANAGEMENT
It is important to note that much of this Dystonia Care Pathway is based on expert opinion, as the evidence for dystonia management in CP is currently limited.

- Rehabilitation Strategies: Rehabilitation strategies used by physiotherapists, occupational therapists and speech pathologists are generally considered cornerstones in the management of dystonia in CP. General principles include: 1) ensuring therapy is goal-directed, 2) avoiding asymmetry and aiming for symmetrical positioning to enhance motor control, 3) optimizing seating and positioning with good stability/support, 4) considering orthoses and splints to increase stability and coordination, and 5) considering the need for communication supports.

- Generalized Dystonia Management: With increasing severity of dystonia, additional interventions may be required beginning with oral medications. Oral baclofen is considered a first line medication for the management of dystonia in CP. Common indications include pain or difficulty sleeping associated with dystonia in CP. If the individual does not respond well to oral baclofen, trihexyphenidyl can be used as a second line medication. Other oral medications should be considered for specific indications. For example, the intermittent use of benzodiazepines is helpful for dystonic storms or disturbed sleep, and gabapentin for dystonia associated with pain.
Clonidine can also be considered for disturbed sleep associated with dystonia.

In the presence of severe generalized dystonia associated with significant impact on care/comfort, more aggressive management can be undertaken. This may include intrathecal baclofen (ITB) or deep brain stimulation (DBS). These strategies require a referral to a specialist team. Individuals with severe generalized hypertonia with a combination of dystonia and spasticity may trial intrathecal baclofen. Other considerations of whether to choose ITB or DBS can be based on the cerebral anatomy and whether placement of the stimulator in the globus pallidus is possible. ITB should be used with caution in the presence of nocturnal respiratory compromise.

A classic feature of dystonia in CP is a fluctuation in the severity of the dystonia. For this reason it is important to periodically re-evaluate the individual and adjust the interventions as required. For some individuals with severe dystonia, a personalized plan for managing increasing dystonia can be developed and includes increasing the dose of current oral medications or introducing a second medication (e.g. clonidine or gabapentin). The use of the Dystonia Severity Action Plan (DSAP) may be helpful for monitoring unstable dystonia and is outlined in Section 3. A rapid and severe increase in dystonia is termed ‘Periodic Status Dystonicus’. It can be life-threatening and requires urgent treatment often with a combination of benzodiazepines and clonidine (enteral, intravenous, or transdermal) (see Section 3 for a management protocol for ‘Status Dystonicus’). Another important issue is the triggering of dystonia from secondary health conditions including gastrointestinal disorders such as reflux or constipation. The overall general health of the individual should be carefully monitored and secondary health issues actively addressed.

**Focal Dystonia Management:** For individuals with focal or segmental dystonia associated with persisting postures causing pain or impacting on function/care-giving, periodic injections of Botulinum toxin can be undertaken. Consider targeting both the agonist/antagonist muscles around the joint(s) involved with the dystonic posture.
The purpose of this document is to provide health care professionals with key facts and recommendations for the assessment and treatment of dystonia in children and youth with cerebral palsy. This summary was produced by the AACPDM Dystonia Care Pathway Team (D Fehlings (team lead), L Brown, A Harvey, K Himmelmann, JP Lin, A MacIntosh, J Mink, E Monbaliu, J Rice, J Silver, L Switzer, I Walters). The summary is based on a systematic review being submitted for peer-reviewed publication. However, health care professionals should continue to use their own judgement and take into account additional relevant factors and context. The AACPDM is not liable for any damages, claims, liabilities, or costs arising from the use of these recommendations including loss or damages arising from any claims made by a third party.