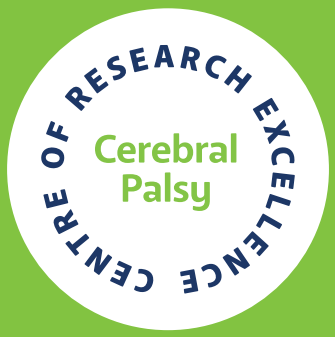

THE IDENTIFICATION AND MEASUREMENT OF DYSKINESIA IN CHILDREN WITH CEREBRAL PALSY

A TOOLKIT FOR CLINICIANS

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CENTRE OF RESEARCH EXCELLENCE IN CEREBRAL PALSY

The CRE-CP is a five-year project funded by the National Health and Medical Research Council that aims to improve the health and well-being of all people affected by cerebral palsy and their families. Leading researchers, clinicians and allied health professionals are joining forces with parents, carers and persons with cerebral palsy in a concerted effort to bring about change in the management and treatment of cerebral palsy. The CRE-CP is a collaboration between a number of highly regarded institutions. The official partners are:

murdoch children's research institute · The Royal Children's Hospital Melbourne · The University of Melbourne · ACU Australian Catholic University · The University of Sydney · University of Notre Dame

Cerebral Palsy Alliance · Telethon Kids Institute · Deakin Worldly · Government of Western Australia Department of Health Child and Adolescent Health Service · CanChild Centre for Childhood Disability Research · ndis National Disability Insurance Scheme · University of Salford Manchester

CONTENTS

1. INTRODUCTION

Dyskinetic cerebral palsy, one of the most disabling forms of cerebral palsy[1], is a motor disorder characterised by changes in muscle tone and posture, with a varying element of involuntary movement[2,3]. Standardised and accurate measurement of dyskinetic cerebral palsy is important to determine intervention effectiveness, ensure our practice is based on high quality evidence and guide future interventions.

The following toolkit summarises the currently available tools that can be utilised to identify and classify dyskinesia in children with cerebral palsy and measure its severity and impact on activity and participation using the framework of the International Classification of Functioning, Disability and Health (ICF)[4]. The implementation of the toolkit aims to increase clinicians' awareness of dyskinesia in children with cerebral palsy, highlight the importance of correct identification and uniform measurement and promote understanding of the impact on treatment interventions selected for these children. The toolkit has been developed as part of a knowledge translation fellowship through the Centre of Research Excellence in Cerebral Palsy (CRE-CP).

The toolkit is not intended as an all-inclusive guide to management of children with dyskinetic cerebral palsy. The toolkit has been developed as a guide for clinicians working with children with dyskinetic cerebral palsy to provide information on current definitions, classification systems, identification and measurement tools.

The CRE-CP is a five-year project funded by the National Health and Medical Research Council, Australia's peak funding body for medical research. The CRE-CP aims to improve the health and wellbeing of all people affected by cerebral palsy and their families. The project brings together leading researchers, clinicians and consumers in a concerted effort to bring about change in the management and treatment of cerebral palsy. The CRE-CP aims to deliver a range of educational materials that will provide thorough information for parents and caregivers, evidence-informed best practice guidelines for clinicians and health professionals, training and education events to disseminate any research findings, a fundamental surveillance system to facilitate timely assessments and interventions, and a sound knowledge translation program to ensure new treatments and management strategies are taken up into routine practice.

2. CLASSIFICATION AND IDENTIFICATION OF DYSKINETIC CEREBRAL PALSY

Cerebral palsy (CP) is the most common cause of motor disability in children, with various international cerebral palsy registers suggesting a prevalence of approximately 2–3 per 1,000 live births[5,6]. Cerebral palsy is an umbrella term that 'describes a group of permanent disorders of the development of movement and posture, causing activity limitation that are attributed to non-progressive disturbances occurring in the developing foetal or infant brain. Its motor disorders are often accompanied by disturbances of sensation, perception, cognition, communication and behaviour, by epilepsy and by secondary musculoskeletal problems'[7]. Cerebral palsy is further classified by its distribution and predominant tone and movement abnormality. Distribution refers to limb involvement, being unilateral or bilateral[5], tone abnormality being hypertonia or hypotonia[7] and movement abnormality or motor type as spastic, dyskinetic (dystonic or choreo-athetoid), ataxic or mixed[5]. Classification of predominant motor type is important for guiding intervention for children with cerebral palsy although it is likely that the majority of children with cerebral palsy present with mixed hypertonia, with components of spasticity, dystonia, choreoathetosis or ataxia[7,8].

2.1 FUNCTIONAL CLASSIFICATION SYSTEMS

Topographical and motor-type classifications are limited as they do not provide information about function, activity or participation or how severely each child is affected. Functional classifications systems, initially developed by Pallisano et al[9] in 1997, provide broad insight into the functioning level of the child and how they mobilise, use their hands, communicate, eat and drink. The functional classification systems don't provide information on the quality of a child's movement but combined with topographical and motor-type classification provide an international language to describe children with cerebral palsy.

The classification systems include:

- Gross Motor Function Classification System (GMFCS)[9,10]
- Manual Ability Classification System (MACS)[11,12]
- Bimanual Fine Motor Function Classification System (BFMF)[13]
- Communication Function Classification System (CFCS)[14]
- Eating and Drinking Ability Classification System (EDACS)[15]

All systems classify children across five levels with Level I indicating minimal disability and a high level of independence and Level V indicating total dependence on equipment and carers for all daily needs. The small number of population based studies of children with dyskinetic cerebral palsy[1,16] have found the majority of these children are more severely impaired than those with bilateral spastic cerebral palsy, functioning at GMFCS and BFMF levels IV and V. It was also found that increasing motor impairment was accompanied by learning disabilities, vision and hearing impairment and epilepsy[16].

GROSS MOTOR FUNCTION CLASSIFICATION SYSTEM (GMFCS – E&R) <2-18 YEARS[9,10]

www.canchild.ca

The GMFCS classifies usual performance, self-initiated gross motor function, such as sitting, crawling, walking and the use of mobility devices. The GMFCS-E&R contains five age bands: Under 2 years; 2–4 years; 4–6 years; 6–12 years and 12–18 years. GMFCS classification should be reassessed after 2 years of age as approximately 40% of children change classification levels by the age of 2[17]. The GMFCS is valid, reliable and predictive.

LEVEL I: Walks without limitations. Able to walk independently on all surfaces. Can run and jump but speed, balance and coordination are reduced.

LEVEL II: Walks with limitations. Able to walk independently but with difficulty on uneven surfaces, inclines and in crowds. Climb stairs using a rail with limited ability (at best) in running and jumping.

LEVEL III: Walks using a hand-held mobility device. Requires walking frame and wheelchair for longer distances. Able to sit independently and independent in floor mobility.

LEVEL IV: Self-mobility with limitations, may use powered mobility. Mobility very limited, requires a wheelchair at home and in the community. May be independent with powered wheelchair. Able to do standing transfers with assistance and requires some support to sit.

LEVEL V: Transported in manual wheelchair. No independent mobility. Requires a carer-pushed wheelchair with seating system.

MANUAL ABILITY CLASSIFICATION SYSTEM (MACS) 4–18 YEARS[11]

www.macs.nu

The MACS classifies typical manual performance, how a child handles objects in daily life irrespective of the differences in function between the two hands[11]. It is not intended to classify best capacity but rather usual performance. It has been found to be valid and reliable[11,18], stable over time[19] and has been used extensively in research and clinical practice.

LEVEL I: Handles objects easily and successfully. At most has limitations in the ease of performing manual tasks requiring speed and accuracy. However, any limitations do not restrict independence in daily living.

LEVEL II: Handles most objects but with somewhat reduced quality and/or speed of achievement. Certain activities may be avoided or achieved with some difficulty. Alternative ways of performance may be used but manual abilities do not usually restrict independence in daily activities.

LEVEL III: Handles objects with difficulty, needs help to prepare and/or modify activities. Performance is slow and achieved with limited success regarding quality and quantity. Activities are performed independently if they have been set up or adapted.

LEVEL IV: Handles a limited selection of easily managed objects in adapted situations. Performs parts of activities with effort and with limited success. Requires continuous support and assistance and/or adapted equipment, for even partial achievement of the activity.

LEVEL V: Does not handle objects and has severely limited ability to perform even simple actions. Requires total assistance.

MINI MANUAL ABILITY CLASSIFICATION SYSTEM (MINI-MACS) 1-4 YEARS[12]

www.macs.nu

The Mini-MACS is an adaption of the MACS for children aged 1– 4 years. It classifies young children's ability to handle objects, appropriate for their age and development. The five levels are similar to those for the MACS.

COMMUNICATION FUNCTION CLASSIFICATION SYSTEM (CFCS) 2-18 YEARS[14]

http://cfcs.us

The CFCS classifies everyday communication performance. It classified communication effectiveness from both a Sender and Receiver perspective, considers familiarity of communication partners and methods of communication e.g. speech, gestures, facial expression and augmentative and alternative communication. It is a valid and reliable classification system[14].

LEVEL I: Effective Sender and Receiver with unfamiliar and familiar partners. Independently alternates between Sender and Receiver roles with most people in most environments. Communication occurs easily and at comfortable pace with unfamiliar and familiar partners. Communication misunderstandings are quickly repaired and do not interfere with overall effectiveness of the person's communication.

LEVEL II: Effective but slower paced Sender and/or Receiver with familiar and unfamiliar partners. Independently alternates between Sender and Receiver roles with most people in most environments, but conversation pace is slow and may make conversation interaction more difficult. May need extra time to understand and compose messages and/or repair misunderstandings. Communication misunderstandings are often repaired and do not interfere with eventual effectiveness of the person's communication with unfamiliar and familiar partners.

LEVEL III: Effective Sender and Receiver with familiar partners. Alternates between Sender and Receiver roles with familiar (but not unfamiliar) conversational partners in most environments. Communication not consistently effective with unfamiliar partners, but usually effective with familiar partners.

LEVEL IV: Inconsistent Sender and/or Receiver with familiar partners. Person doesn't consistently alternate Sender and Receiver roles. May: a) occasionally be effective Sender and Receiver; b) be effective Sender but limited Receiver; c) be limited Sender but effective Receiver. Communication sometimes effective with familiar partners.

LEVEL V: Seldom effective Sender and Receiver even with familiar partners. Limited as Sender and Receiver. Communication difficult for most people to understand. Appears to have limited understanding of messages from most people. Communication seldom effective even with familiar partners.

BIMANUAL FINE MOTOR FUNCTION CLASSIFICATION SYSTEM (BFMF)[13]

The BFMF describes fine motor function by classifying the ability to grasp, hold and manipulate objects in each hand separately[13]. The BFMF has been found to be valid and complements the MACS by providing classification of fine motor function and actual use of the hands[20]. The BFMF is the main classification system utilised within the Surveillance of Cerebral Palsy in Europe register.

LEVEL I: One hand manipulates without restrictions. The other hand manipulates without restrictions or has limitations in more advanced fine motor skills.

LEVEL II:

(a) One hand manipulates without restrictions. The other hand has the ability to grasp or hold.

(b) Both hands have limitations in more advanced fine motor skills.

LEVEL III: The child needs help with tasks.

(a) One hand manipulates without restrictions. The other hand has no functional ability.

(b) One hand has limitations in more advanced fine motor skills. The other hand has only ability to grasp or worse.

LEVEL IV: The child needs support and/or adapted equipment.

(a) Both hands have only ability to grasp.

(b) One hand has only ability to grasp. The other hand has only ability to hold or worse.

LEVEL V: The child requires total assistance, even with adaptations.

Both hands have only ability to hold or worse.

EATING AND DRINKING ABILITY CLASSIFICATION SYSTEM (EDACS)[15]

www.sussexcommunity.nhs.uk

The EDACS classified a child's usual ability to eat and drink with consideration of efficiency, safety and the level of assistance required.

LEVEL I: Eats and drinks safely and efficiently.

LEVEL II: Eats and drinks safely but with some limitations to efficiency.

LEVEL III: Eats and drinks with some limitations to safety, maybe limitations to efficiency.

LEVEL IV: Eats and drinks with significant limitations to safety.

LEVEL V: Unable to eat or drink safely, tube feeding may be considered to provide nutrition.

Level of assistance required:

INDEPENDENT (IND): Able to eat and drink without any assistance.

REQUIRES ASSISTANCE (RA): Requires help to bring food or drink to mouth, either from another person or through use of adapted equipment.

TOTALLY DEPENDENT (TD): Totally dependent on another person to bring food and drink to mouth.

In addition to the classification systems the Functional Mobility Scale (FMS)[21] classifies a child's functional mobility, taking into account the range of assistive mobility devices they use.

FUNCTIONAL MOBILITY SCALE (FMS)[21]

www.rch.org.au

The FMS classifies a child's functional mobility, with and without assistive devices, across a range of settings. It takes into account passive mobility, assisted movement and self-initiated mobility. It rates mobility for three specific distances: 5m, 50m and 500m representing the home, school and community settings. The FMS is a reliable tool that is able to detect changes in mobility following intervention.

RATING 1: Uses wheelchair.

RATING 2: Uses walker without help.

RATING 3: Uses crutches without help.

RATING 4: Uses sticks (one or two) without help.

RATING 5: Independent on level surfaces, requires rail for stairs.

RATING 6: Independent on all surfaces.

2.2 INCIDENCE AND REPORTING OF DYSKINETIC CEREBRAL PALSY

There appears some inconsistency in the identification and reporting of dyskinesia in children with cerebral palsy between clinicians (see survey results, Appendix 2), the different international cerebral palsy registers[16,22] and in the literature[5,7.] This is most likely due to clinical under recognition of the various movement disorders[23] despite a more recent increase in the understanding and definition of dyskinetic cerebral palsy as well as the complexity of discriminating between the different movement disorders themselves when they are frequently found in combination[24]. A study looking at trends and prevalence of dyskinetic cerebral palsy across Europe[16] found the incidence of dyskinetic cerebral palsy appears to be increasing. Population-based data from various international cerebral palsy registers suggests prevalence of dyskinetic cerebral palsy varies[1].
The Surveillance of Cerebral Palsy in Europe (SCPE), using data from eight centres, reported a rate of 14.4%[25] similar to the rate of 15% reported in Sweden[1]. The rate in Australia appears lower, with dyskinetic cerebral palsy comprising 5.9% and ataxic cerebral palsy 5.3%[26].
The differences reported from various registers may relate to under identification of dyskinetic cerebral palsy in some countries and also to how predominant motor types are designated.

There has been increasing interest and focus on dystonia and dyskinetic cerebral palsy internationally and in the literature. There is a move to more accurately describe cerebral palsy by the predominant motor type[7], hence the greater focus on dystonic cerebral palsy and dyskinetic cerebral palsy. The new Care Pathway on Dystonia in Cerebral Palsy developed by the American Academy for Cerebral Palsy and Developmental Medicine (AACPDM, 2016) (**https://www.aacpdm.org/publications/care-pathways/dystonia**) provides evidence informed guidelines for clinicians about dystonia in cerebral palsy, its identification and management. Accurate identification and classification is imperative to guide both medical and therapy intervention as children with dyskinetic cerebral palsy can have different outcomes from the same treatments to those children with spasticity (which is more commonly seen and understood in cerebral palsy). Standardised and accurate measurement of dyskinesia in cerebral palsy is important to determine intervention effectiveness, ensure our practice is based on high quality evidence and guide future interventions.

2.3 DEFINITIONS

ATHETOSIS: is 'a slow, continuous, involuntary writhing movement that prevents maintenance of stable posture'[27]. Movements appear smooth and random and usually involve the distal extremities more than proximal body areas. Athetosis generally appears in combination with dystonia and chorea and choreoathetosis is often caused by dyskinetic cerebral palsy in combination with dystonia.

CHOREA: is an 'ongoing random-appearing sequence of one or more discrete involuntary movements or movement fragments'[27]. It is distinguished from dystonia by its unpredictable, random, rapid and continuous nature. Chorea does not appear to be linked to voluntary movement and movement does not cease on relaxation so children tend to present in constant motion.

DYSKINETIC CEREBRAL PALSY: is defined, according to the SCPE[5] as 'involuntary, uncontrolled, recurring and occasionally stereotyped movements. Primitive reflex patterns predominate and muscle tone varies'. This is further sub-grouped into dystonic and choreoathetotic. The term dyskinetic cerebral palsy tends to be used when the dominance of dystonia and choreoathetosis is difficult to delineate[28].

DYSTONIA: an involuntary alteration in muscle activation patterns during voluntary movement or the maintenance of posture. It is a 'movement disorder in which involuntary sustained or intermittent muscle contractions cause twisting or repetitive movements, abnormal postures, or both'[8]. 'Dystonia is a movement disorder characterised by sustained or intermittent muscle contractions causing abnormal, often repetitive movements, postures or both. Dystonic movements are typically patterned, twisting, and may be tremulous. Dystonia is often initiated or worsened by voluntary action and associated with overflow muscle activation'[29].

HYPERKINETIC MOVEMENT DISORDERS: are defined as any unwanted excess movement[27]. They are characterised by excessive involuntary movements including dystonia, chorea, athetosis, and myoclonus arising from many aetiologies including congenital, acquired, neurodegenerative, and genetic disorders. The most common cause of hyperkinetic movement disorders in children is dyskinetic cerebral palsy[27].

HYPERTONIA: 'abnormally increased resistance to externally imposed movement about a joint'[8] that may be caused by spasticity, dystonia, rigidity or a combination of these.

HYPOTONIA: Hypotonic cerebral palsy is characterised by generalised muscular hypotonia that persists beyond 3 years of age and does not result from a primary disorder of muscle or peripheral nerves[30].

MIXED CEREBRAL PALSY: this term is frequently found in the literature and used clinically when describing patients who present with a mixed pattern of hypertonias, or a combination of spasticity and dystonia. Unfortunately, it does not give an indication of which hypertonia is dominant or where each type of hypertonia predominantly occurs, for example spasticity affecting the lower limbs and dystonia the upper limbs.

MYOCLONUS: is a sequence of repeated, frequently non-rhythmic, brief shock-like jerks caused by sudden involuntary contraction (positive myoclonus) or relaxation (negative myoclonus) of one or more muscles[27]. Myoclonus can be caused by or worsened by movement.

SPASTICITY: a velocity-dependent resistance to muscle stretch that occurs when 'resistance to externally imposed movement increases with increasing speed of stretch and varies with the direction of joint movement' and/or 'resistance to externally imposed movement rises rapidly above a threshold speed or joint angle'[8].

TONE: 'resistance to stretch while patient is attempting to maintain a relaxed state of muscle activity'[8]. It can be described as increased or decreased at rest and excludes resistance that results from joint contracture. Tone is assessed clinically to determine muscle resistance by passive joint movement.

3. DYSKINESIA SCREENING TOOLS FOR CHILDREN WITH CEREBRAL PALSY

The accurate differentiation of hypertonia in children with cerebral palsy ensures not only accurate classification of motor types but helps maximise intervention outcomes, as response to treatment can vary depending on predominant motor type. The identification and differentiation of movement disorders is generally completed by an experienced examiner during a neurological examination[31]. In 2010 the Hypertonia Assessment Tool (HAT) was published and can now also be utilised to reliably differentiate between paediatric hypertonia sub-types[31]. A tool to accurately identify choreoathetosis is still lacking.

3.1 HYPERTONIA ASSESSMENT TOOL (HAT)

The Hypertonia Assessment Tool[31], a standardised tool that accurately identifies the presence of spasticity, dystonia and rigidity in people with cerebral palsy, was developed to address the need for clinical differentiation of hypertonias: spasticity, dystonia and rigidity. It does not identify the presence or absence of choreoathetosis.

The HAT is a seven item clinical assessment tool developed for use with children aged 4–19 years to differentiate paediatric hypertonia sub-types (Figure 1). The child's limbs are touched or moved in a series of movements to illicit movement, increased tone and/or resistance. The tool consists of two items for spasticity, two for rigidity and three for dystonia. The items are all scored as either present or absent indicating the presence or absence of that hypertonia sub-type. Mixed hypertonia is indicated if two or more items from the three sub-types are scored as present. The HAT has good reliability and validity for the identification of spasticity and the absence of rigidity and moderate reliability and validity findings for dystonia[31]. Videotape review is not necessary to improve scoring of the HAT items[32]. The manual and score sheets can be downloaded from the website: **https://research.hollandbloorview.ca/outcomemeasures/hat**

Clinical utility: The HAT has good clinical utility and is accompanied by a downloadable user manual with clear assessment and scoring instruction. The benefit of the HAT is its ability to be applied by all clinicians to distinguish between spasticity and dystonia in children with cerebral palsy rather than neurological examination requiring an experienced practitioner. It can be used in the clinical setting to discriminate between the different hypertonias to specifically target, for example spasticity and or dystonia and improve treatment outcomes and to help clarify study outcomes in the research setting by classification of participant hypertonia sub-types.

Figure 1: Hypertonia Assessment Tool (HAT) – Scoring Chart

Holland Bloorview Kids Rehabilitation Hospital

Bloorview RESEARCH INSTITUTE

HYPERTONIA ASSESSMENT TOOL (HAT) - SCORING CHART

Name:_____ **Chart/File #:**_____

Clinical Diagnosis:_____ **Date of Birth:** _____

Limb Assessed: **Gender:** ☐ Male ☐ Female

☐ Arm ☐ Left ☐ Right **HAT Assessor:**_____

☐ Leg ☐ Left ☐ Right **Date of Assessment:** _____

HYPERTONIA ASSESSMENT TOOL (HAT)

HAT ITEM	SCORING GUIDELINES (0=negative or 1=positive)	SCORE 0=negative 1=positive *(circle score)*	TYPE OF HYPERTONIA
1. Increased involuntary movements/postures of the designated limb with tactile stimulus of another body part	0= No involuntary movements or postures observed	0	DYSTONIA
	1= Involuntary movements or postures observed	1	
2. Increased involuntary movements/postures with purposeful movements of another body part	0= No involuntary movements or postures observed	0	DYSTONIA
	1= Involuntary movements or postures observed	1	
3. Velocity dependent resistance to stretch	0= No increased resistance noticed during fast stretch compared to slow stretch	0	SPASTICITY
	1= Increased resistance noticed during fast stretch compared to slow stretch	1	
4. Presence of a spastic catch	0= No spastic catch noted	0	SPASTICITY
	1= Spastic catch noted	1	
5. Equal resistance to passive stretch during bi-directional movement of a joint	0= Equal resistance not noted with bi-directional movement	0	RIGIDITY
	1= Equal resistance noted with bi-directional movement	1	
6. Increased tone with movement of another body part	0= No increased tone noted with purposeful movement	0	DYSTONIA
	1= Greater tone noted with purposeful movement	1	
7. Maintenance of limb position after passive movement	0= Limb returns (partially or fully) to original position	0	RIGIDITY
	1= Limb remains in final position of stretch	1	

SUMMARY SCORE – HAT DIAGNOSIS

		Check box:
DYSTONIA →	Positive score (1) on at least one of the Items #1, 2, or 6	☐ Yes ☐ No
SPASTICITY →	Positive score (1) on either one or both of the Items #3 or 4	☐ Yes ☐ No
RIGIDITY →	Positive score (1) on either one or both of the Items #5 or 7	☐ Yes ☐ No
MIXED TONE →	Presence of 1 or more subgroups (e.g. dystonia, spasticity, rigidity)	☐ Yes ☐ No

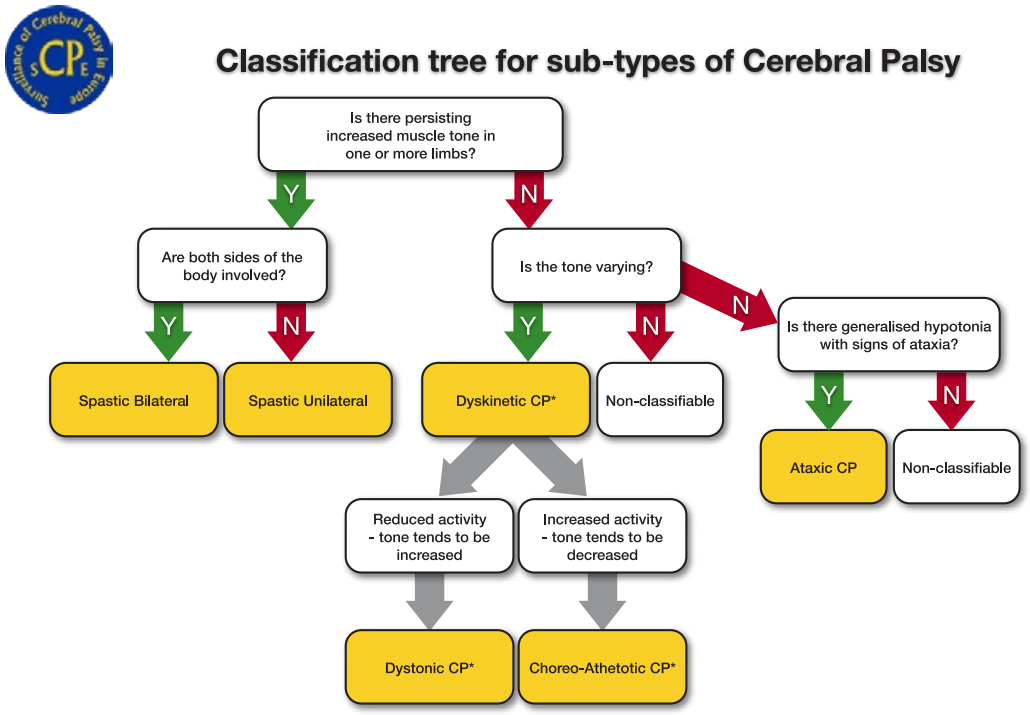
HAT DIAGNOSIS: *(Fill in all that apply)* _____

HAT Manual can be accessed at http://www.hollandbloorview.ca/research/scientistprofiles/fehlings.php

4. CLASSIFICATION OF DYSKINETIC CEREBRAL PALSY

Classification of predominant motor types is reported clinically and addressed in the various international cerebral palsy registers. In Europe, classification of the predominant motor type is completed using the Surveillance of Cerebral Palsy in Europe (SCPE) hierarchical classification tree of cerebral palsy sub-types[5]. This classification relies on clinical judgement and has the primary purpose of standardising classification for the monitoring of trends in rates of cerebral palsy across Europe[5]. SCPE classifies cerebral palsy into three main groups based on neurological signs: spastic, ataxic and dyskinetic (dystonic, choreo and athetoid) cerebral palsy (Figure 2). Those children with mixed motor sub-types are classified according to their predominant clinical feature[28]. One limitation of this hierarchical classification is 'mixed' tone cannot be easily accounted for, only the predominant motor type.

Figure 2: Surveillance of Cerebral Palsy in Europe (SCPE) hierarchical classification tree of cerebral palsy sub-types

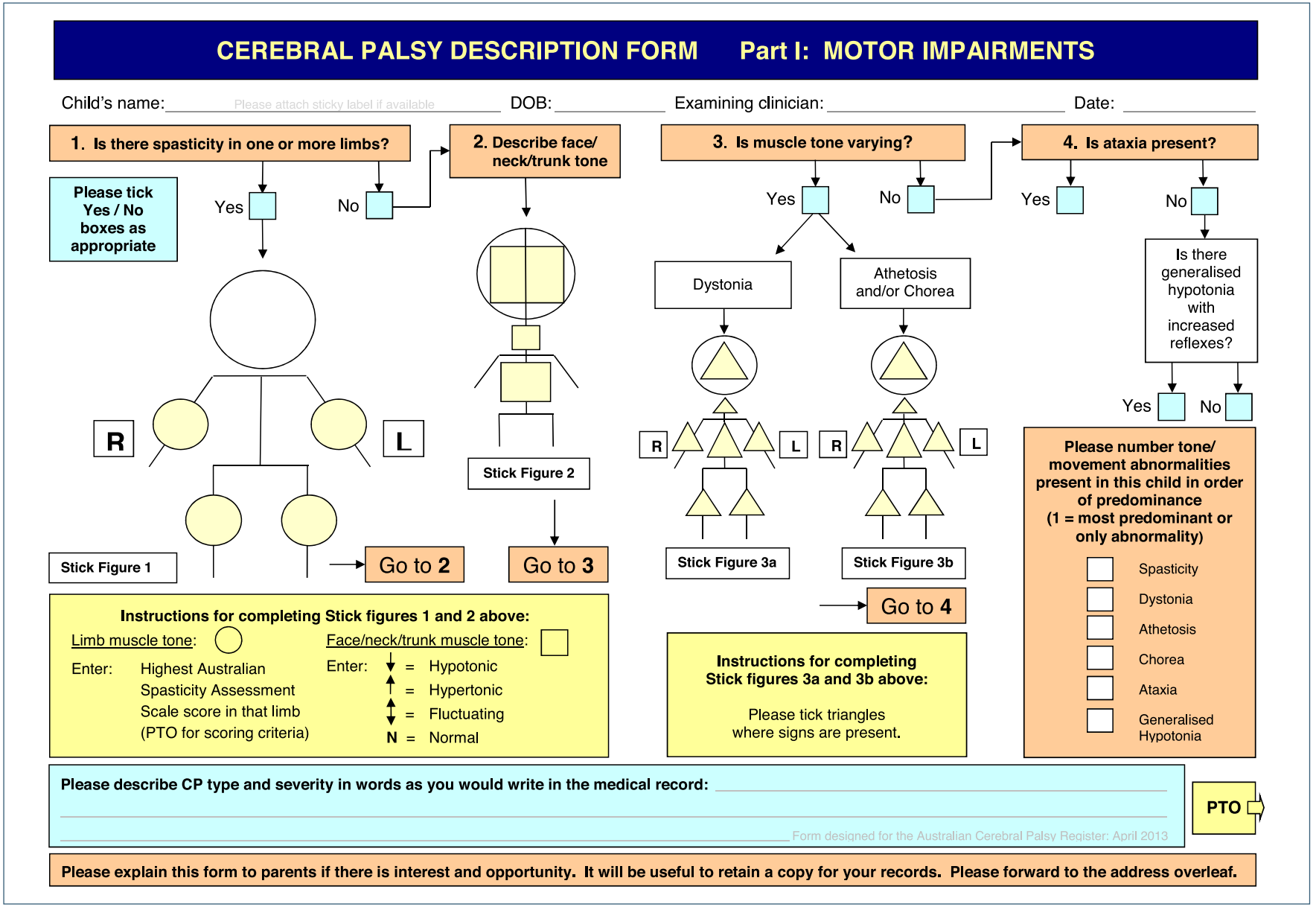


SCPE Collaborative Group. Surveillance of cerebral palsy in Europe: a collaboration of cerebral palsy surveys and registers. *Developmental Medicine and Child Neurology.* 2000;42:816-24.

Predominant motor sub-type is also used for classification purposes in the Australian Cerebral Palsy Registers (ACPR)[33,34]. The ACPR utilise the Cerebral Palsy Description Form: Motor Impairments (Figure 3). This form can be used in combination with the Australian Spasticity Assessment Scale (ASAS)[35]. This form applies a limb by limb approach to provide an objective clinical picture of the child with cerebral palsy[35], coding spasticity using the ASAS as well as identifying anatomical distribution of dystonia and athetosis. The form enables ranking of predominant motor types with the possibility for equal rankings[35]. It is widely recognised that many children with cerebral palsy have both spastic and dyskinetic features[8] and the general consensus is to continue to classify by dominant motor type but in addition list secondary motor types and the term 'mixed' should be accompanied by elaboration of the component motor disorders[7,8].

Figure 3: Cerebral Palsy Description Form - Motor Impairments (Australian Cerebral Palsy Register)

http://ww2.health.wa.gov.au/~/media/Files/Corporate/general%20documents/communicable%20diseases/PDF/Cerebral_Palsy_Description_Form_WARDA_website.ashx



5. CLASSIFICATION OF DYSTONIA

Dystonia has historically been classified by aetiology into two broad categories:

1a. Primary dystonia: children with primary dystonias have no structural brain abnormality, often no underlying cause and may be genetically based. Dystonia is the only neurological feature.

1b. Primary-plus dystonia: children with primary-plus dystonias present with additional movement disorders such as myoclonus dystonia.

2. Secondary dystonia: children with secondary dystonias have structural abnormalities of the brain caused by damage or degeneration to the brain. This can be further subdivided into those arising from a static injury to the brain, including children with acquired brain injuries and cerebral palsy, and those caused by an underlying progressive condition.

Albanese and colleagues (2013)[29] identifying inconsistencies with this system of classification, including the terminology, proposed a new classification system based on aetiology, age at onset and body distribution.

The proposed two categories are:

1. AXIS 1
The first axis describes the clinical characteristics including age of onset (infancy, childhood, adolescents, early and late adulthood), body distribution (focal, segmental, multifocal, generalised, hemi dystonia), temporal pattern (static or progressive and persistent, action-specific, diurnal or paroxysmal) and associated features (isolated or combined with another movement disorder and other neurological manifestations).

2. AXIS 2
The second axis addresses aetiology. This includes nervous system pathology (evidence or not of degeneration or structural lesions) and whether the dystonia is inherited (proven genetic origin), acquired (due to known specific cause) or idiopathic (unknown cause).

Children with cerebral palsy would generally be classified as presenting with static, generalised, infant or childhood onset dystonia, frequently combined with other movement disorder/s, with evidence of structural lesions due to cerebral palsy.

6. OUTCOME MEASURES FOR CHILDREN WITH DYSKINETIC CEREBRAL PALSY

The presence of dystonia and/or choreoathetosis in children with cerebral palsy impacts on function and it is recognised that dyskinetic cerebral palsy is one of the most disabling forms of cerebral palsy in children[1]. Accurate identification of motor sub-types is important for guiding intervention as different movement disorders can respond in varying ways to the same intervention. Assessment of the severity of the dystonia and choreoathetosis assists the clinician in objectively quantifying the movement disorder as well as monitoring intervention outcomes. One of the clinical features of dyskinetic cerebral palsy is the fluctuation in its severity due to mood, general health, mental health and environmental influences. This can make reliable assessment of severity difficult.

The ideal dystonia/choreoathetosis scale should be concise and simple to administer and score, both in the clinical and research setting. Scales should have the ability to rate current presentation as well as improvement or deterioration and provide some information about the severity of dystonia/choreoathetosis at rest and on activity and the influence of environmental or health impacts. Scale items must be clearly defined to ensure high inter-rater reliability. Ideally a scale should be able to discriminate between different movement disorder sub-types and specifically quantify the effects of interventions. In addition to impairment severity, scales should address areas deemed important to patients and their families including impact on activities of daily living, burden of care and wellbeing.

A number of rating scales have been developed for use with patients with primary and secondary dystonia and/or choreoathetosis. The published scales that have been utilised specifically with children with cerebral palsy[36] include:

- Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS)[37]
- Barry Albright Dystonia Scale (BADS)[38]
- Unified Dystonia Rating Scale (UDRS)[39]
- Movement Disorder Childhood Rating Scale (MD-CRS)[40]
- Movement Disorder Childhood Rating Scale 0-3 (MD-CRS 0-3)[41]
- Dyskinesia Impairment Scale (DIS)[42]

The majority of these scales assess dystonia severity whilst the most recently published DIS is the only scale to address both dystonia and choreoathetosis in cerebral palsy. The MD-CRS and MD-CRS (0-3) purport to measure chorea and athetosis but lack definitions of these movement disorders and were not specifically designed for children with cerebral palsy or secondary movement disorders. All scales are intended as impairment assessments at the Body Functions and Structures level according to the ICF,[4] with some of the scales providing select insight into the impact of the dystonia or movement disorder on activity. All the scales assess dystonia and/or choreoathetosis at the eyes, mouth (and impact on speech), neck, trunk upper and lower limbs with the UDRS and DIS being the only two to discriminate between proximal and distal aspects of the limbs. This may have functional implications, as a study by Monbaliu et al (2016)[43] found proximal limb dystonia to have greater impact on functional activity ability than distal limb dystonia.

The scales are presented, as follows, in order of their publication. Sample scoring sheets for each scale are contained in Appendix 1.

6.1 BURKE-FAHN-MARSDEN DYSTONIA RATING SCALE (BFMDRS)

DESCRIPTION: The BFMDRS[37] was developed to assess adults with primary torsion dystonia. It was the first scale to specifically assess generalised dystonia as a complex movement disorder. There are two sections to the tool, a Movement Scale based on physical examination and a Disability Scale based on the patient's view of the impact of their dystonia on activities of daily living. The Movement Scale evaluates dystonia across nine body regions using a 'severity rating' to quantify the dystonia and a 'provoking factor' for when dystonia occurs. The Disability Scale rates how dystonia affects select activities.

PSYCHOMETRIC PROPERTIES: The BFMDRS has been examined in multiple studies involving children with cerebral palsy. These demonstrate moderate concurrent[44,45] and limited predictive[46] validity and moderate expert opinion for content validity[44,47,48] according to the Consensus-based Standards for selection of health Measurement Instruments (COSMIN)[49]. There is moderate evidence for its internal consistency and inter-rater reliability[44] and no evidence for intra-rater or test-retest reliability. Eleven intervention studies, specifically involving children with cerebral palsy, indicate the Movement Scale has excellent responsiveness to change but this is less so for the Disability Scale. Responsiveness to change has predominantly been assessed in studies investigating deep brain stimulation in children with cerebral palsy (n=8)[46-48,50-54] with three further studies investigating intrathecal baclofen (n=1)[45], Botulinum toxin-A (n=1)[55] and a medication trial (n=1)[56].

CLINICAL UTILITY: The BFMDRS is readily available via the original journal article at no cost. No manual or training package is available. Administration instructions are adequate. Scoring criteria are outlined and higher total scores indicate greater severity of dystonia. A video protocol, of less than five minutes duration, is outlined, although scoring time is not stated (see Table 2). No details are provided regarding level of clinical knowledge required for use or interpretation of scores. The BFMDRS was designed for adults with primary dystonia, but has been utilised as an outcome measure in many studies investigating children with cerebral palsy although the authors suggest results should be interpreted with caution when the scale is utilised for people with secondary dystonia[37].

INTENDED POPULATION: Adults with generalised primary dystonia, age not specified.

REPORTING STYLE: Observational and self-report.

ADMINISTRATION: The Movement Scale examination is completed with the patient sitting, standing, walking and engaging in various movements and activities. A detailed assessment protocol is provided in the original article as well as a clear video protocol (Table 1).

Table 1: Video protocol for the BFMDRS

Sitting at rest, arms resting on legs - Whole body - Zoom in to different body regions (head and neck, each hand, trunk, each foot)	45 sec
Speak – name, date, describe speech, swallowing and current problems - Film whole body - Zoom in to different body parts	45 sec
Arms suspended in front of body - Finger to nose x 5 - Rapid succession movements: each hand (open and close) and foot (tapping)	45 sec
Arise and stand, turn 90˚ x4	30 sec
Walk - Whole body - Zoom in to different body regions	60 sec
Write with each hand: name, date, sentence, spiral - Whole body - Zoom in activity	30 sec
Total	**4 minutes 15 seconds**

SCORING: The BFMDRS has two sections:

Movement Scale evaluates dystonia on a five-point scale across nine body regions: eyes, mouth, speech and swallow, neck, trunk, right and left upper limb, and right and left lower limb. It is scored using a 'severity rating' to quantify dystonia in each region regardless of the circumstances in which it occurs, and a 'provoking factor' to indicate the situations under which dystonia occurs. Scores for the provoking factor and severity factor for each body region are multiplied together to give a product for that body region. The eyes, mouth and neck regions are further multiplied by 0.5 to 'down weight' these scores as their involvement is argued to add less to overall disability. The maximum score is 120, the higher the score the more severe the dystonia.

Shoulder and Pelvic girdle dystonia is scored as per the following criteria:

- Trapezius dystonia scored as part of neck region
- Shoulder girdle dystonia affecting upper limb placement is scored as part of upper limb region
- Shoulder girdle dystonia that accompanies kyphosis or scoliosis is considered part of the trunk region
- Tortipelvis is scored as part of the trunk region
- Pelvic dystonia affecting lower limb placement is scored as part of the lower limbs

Disability Scale rates the patient's view of the impact of their dystonia on activities of daily living: speech, handwriting, feeding, eating/swallowing, hygiene, dressing and walking. The maximum score for the Disability Scale is 30. See Table 2 for scoring criteria.

Table 2: Scoring criteria for the BFMDRS

MOVEMENT SCALE	SCORE	SCORING CRITERIA
Severity factor	0	No dystonia present
	1	Slight dystonia, clinically insignificant
	2	Mild dystonia, obvious but not disabling
	3	Moderate dystonia that interferes but doesn't prevent function
	4	Severe dystonia that prevents function at that body part
Provoking factor	0	No dystonia during movement
	1	Least severe dystonia where dystonia is seen only on a particular action
	2	Dystonia observed on many actions
	3	Dystonia present on action of distant part or intermittently at rest
	4	Dystonia present at rest, the most severe state of persistent dystonia
DISABILITY SCALE		
	0	Normal
	1	Slight difficulty or abnormality
	2	Some difficulty, requires help with some activities
	3	Marked difficulty, requires help with most activities
	4	Completely dependent
	6	Wheelchair bound (for walking only)

6.2 BARRY ALBRIGHT DYSTONIA SCALE (BADS)

DESCRIPTION: The BADS[38] based on the BFMDRS[37], was developed specifically to assess secondary dystonia in patients with cerebral palsy and acquired brain injury with limitations in cognition and physical ability. Scoring is based on severity of posturing and involuntary dystonic movements rather than on functional ability as the authors hypothesised that the intended population for the BADS may not be expected to gain function following an intervention but rather make improvements in ease of care and comfort. The BADS rates dystonia severity. The BADS does not assess functional tasks and the authors recommend the use of other functional assessment tools such as the Pediatric Evaluation of Disability Inventory (PEDI)[57], Gross Motor Function Measure (GMFM)[58] and the Canadian Occupational Performance Measure (COPM)[59] that have demonstrated reliability and validity for people with cerebral palsy and acquired brain injury.

PSYCHOMETRIC PROPERTIES: The BADS is the most commonly reported measure for studies investigating interventions for children with dyskinetic cerebral palsy, despite it having limited evidence of reliability in this diagnostic group. One study by Monbaliu et al (2010)[44] indicates the BADS has moderate internal consistency and inter-rater reliability. No evidence of intra-rater or test-retest reliability are evident specifically for children with cerebral palsy. Three fair studies provide moderate evidence of construct validity[60,61] with moderate concurrent[42,44,45] and limited predictive validity evident[40]. Expert opinion demonstrates limited content validity[44]. Eleven studies provide excellent evidence of responsiveness to change across multiple interventions including deep brain stimulation (n=3)[48,50,62], intrathecal baclofen (n=5)[38,45,63-65], intra-ventricular baclofen (n=1)[66], baclofen bolus test dose (n=1)[67] and medication RCT (n=1)[68], however data is predominantly descriptive and more specific analysis is warranted.

CLINICAL UTILITY: The BADS appears to have the greatest clinical utility[36] of all the presented scales for children with cerebral palsy and dystonia. It was developed specifically for people with cerebral palsy, is readily available via journal article and quick and easy to administer. No manual is available and although video is suggested no details are provided. The authors mention training improves reliability but details of the training are not provided. Scoring time is not indicated but scoring criteria are clear with explanation for areas unable to be assessed.

INTENDED POPULATION: Patients with secondary dystonia due to cerebral palsy and acquired brain injury, age not specified.

REPORTING STYLE: Observational.

ADMINISTRATION: Video-taping is recommended to enable pre and post intervention comparison. A standardised video script, indicating between 20 to 45 minutes, is mentioned but details are not provided beyond asking patients to remain still then perform a variety of functional tasks, depending on their capability.

SCORING: The BADS scores dystonia on a five-point criterion-based, ordinal severity scale across eight body regions: eyes, mouth, neck, trunk, right and left upper limbs, and right and left lower limbs. A maximum score is 32, with higher scores indicating more severe dystonia and decreased functional ability (see Table 3 for scoring criteria). If a body region is unable to be scored, this body region is excluded and a reduced total score obtained. If the patient is unable to perform even the simple functional tasks, such as sitting in a chair, then the abnormal posturing or muscle contraction severity determine the score.

Table 3: Scoring criteria for BADS

SCORE	SCORING CRITERIA
0	No dystonia
1	Slight dystonia, present less than 10% of the time
2	Mild dystonia, present less than 50% of the time and does not interfere with function
3	Moderate dystonia, present more than 50% of the time and interferes with function
4	Severe dystonia, present more than 50% of the time and prevents function

6.3 UNIFIED DYSTONIA RATING SCALE (UDRS)

DESCRIPTION: The UDRS[39] was developed by the Dystonia Study Group, for use with adults with primary dystonia, in response to perceived limitations they felt were found in the BFMDRS. It includes more detailed assessment of individual body parts and rates the proximal and distal aspects of the upper and lower limbs. It also eliminates subjective patient ratings in the areas of speech and swallow. The scale assesses dystonia severity across 14 body regions using a Duration Factor and Motor Severity Factor. A detailed video protocol, utilised for scoring, accompanies the tool. The UDRS does not assess the influence of dystonia on functional activity, although some functional activities are included in the video protocol, for example walking and writing.

PSYCHOMETRIC PROPERTIES: The UDRS has little available literature regarding its use with children with cerebral palsy, since this was not its published target population. Reliability studies demonstrate moderate internal consistency and inter-rater reliability[44]. Validity studies are limited to expert opinion on content validity[44] and moderate concurrent validity[44]. One intervention trial demonstrated limited evidence for responsiveness to change pre-post Botulinum toxin-A in children with cerebral palsy[55].

CLINICAL UTILITY: The UDRS has adequate clinical utility for children with cerebral palsy despite this not being the intended target population. The scale is available in the original journal article, including adequate instruction and a detailed video protocol. Authors suggest experience improves reliability. Scoring of the Duration Factor sub-scale is complex and knowledge of patients range motion is required prior to scoring. Many of the activities assessed as part of the scale are not applicable to children with more severe presentations of dyskinetic cerebral palsy, for example drawing spirals with both hands. All information gained from the scale is at the Body Functions and Structures level of the ICF with no Function or Participation level insights.

INTENDED POPULATION: Adults with generalised primary dystonia, age not specified.

REPORTING STYLE: Observational and patient interview.

ADMINISTRATION: The UDRS can be scored from video (see Table 4). Patient Swallow interview:

- Do you have problems with swallowing? If yes, is it occasional or frequent?
- Do you choke occasionally or frequently?, Can you swallow firm foods? Liquids?

SCORING: The UDRS scores dystonia severity using a Duration factor and Motor Severity Factor across 14 body regions: eyes and upper face, lower face, jaw and tongue, larynx, neck, trunk, left and right shoulder/proximal arm, left and right distal arm/hand, left and right proximal leg, and left and right distal leg/foot. The Duration Factor rates frequency of the dystonia and if it is predominantly sub maximum or maximum intensity. The Motor Severity Factor quantifies the percentage of range of motion in which the dystonic movements occur (see Table 5 for scoring criteria). Each Factor can score a maximum of 56, giving a scale total score of 112. Higher scores indicate more severe dystonia.

Table 4: UDRS video protocol[39]

AREA ASSESSED	PERSPECTIVE	ACTIVITY	TIME
Eyes and upper face	Close view of head and shoulders sitting unsupported on stool	At rest Eyes open Eyes closed Forced eye blinks x10 Close view face – rest	10 sec 10 sec close/10 sec far 10 sec close/10 sec far 10 sec 10 sec
Lower face, jaw, tongue, larynx	Patient seated	Read: standard passage Repeat: *tee*, *mee*, *la*, *ca* x5 Hold note *eee* Count to 10 Tongue protrusion Open and close mouth x5	 15 sec 5 sec 5 sec 5 sec 10 sec
Neck	Seated in chair, close view head and shoulders	Front view at rest Seated, eyes closed Quiet conversation Turn head to L and R Tilt ears to shoulders Look up and down Lateral view Walk back and forth x2	10 sec 10 sec 10 sec 5 sec 20 sec
Shoulders and upper arms, distal arm and hands	Far view upper half of body	Arms ext supinated Arms ext pronated Arms flexed at elbow Finger to nose x5 Finger tapping, R and L x5 Flex and ext wrists, arms ext x5 Cup to lips, R and L Write '*Today is a nice day*' x3 Draw spirals, without resting hand on paper R and L Hold up spiral	5 sec 5 sec 5 sec 5 sec 5 sec 5 sec 5 sec 15 sec 10 sec
Upper leg, distal leg, foot and trunk	Far view entire body, sitting Far view whole body, standing and walking	Sitting quietly Heel toe taps x5 R and L Standing: frontal view Standing: lateral view Walking: 20 feet x2 reps	10 sec 10 sec 10 sec 5 sec 20 sec

Table 5: Scoring criteria for UDRS

MOVEMENT SCALE	SCORE	SCORING CRITERIA
Duration Factor	0	No duration
	0.5	Occasional dystonia, ≤25% of the time, predominantly submaximal
	1.0	Occasional dystonia, ≤25% of the time, predominantly maximal
	1.5	Intermittent, 25-50% of the time, predominantly submaximal
	2.0	Intermittent, 25-50% of the time, predominantly maximal
	2.5	Frequent, 50-75% of the time, predominantly submaximal
	3.0	Frequent, 50-75% of the time, predominantly maximal
	3.5	Constant, ≥75% of the time, predominantly submaximal
	4.0	Constant, ≥75% of the time, predominantly maximal
Motor Severity Factor	0	No dystonia
	1	Mild dystonia, ≤25% intensity/possible range
	2	Moderate dystonia, >25% and ≤50% intensity/possible range
	3	Severe dystonia, >50% and ≤75% intensity/possible range
	4	Extreme dystonia, >75% intensity/possible range

6.4 MOVEMENT DISORDER – CHILDHOOD RATING SCALE (MD-CRS)

DESCRIPTION: The MD-CRS[40] was developed for children aged four to 18 years to: (i) describe clinical features of different movement disorders; (ii) evaluate the intensity of movement disorders in different body regions at rest and on activity; (iii) assess the influence of movement disorders on motor function and activities of daily living; and (iv) explore the impact of movement disorders on neurodevelopment. The different movement disorders it can assess include: Hypokinetic-rigid, Chorea/Ballism, Dystonia/Athetosis, Myoclonus, Tic, and tremor. These movement disorders are not defined. The scale consists of two parts. Part 1 is a 'General Assessment' and consists of Motor Function, Oral/Verbal Function, Self-Care and Attention/Alertness. Part 2 assesses 'Movement Disorder Severity' across seven body regions.

PSYCHOMETRIC PROPERTIES: No articles reporting on the validity or reliability of the MD-CRS specifically in children with cerebral palsy are available. One medication trial study[69] indicates the tool is suitable to detect change following an intervention in children with cerebral palsy.

CLINICAL UTILITY: The MD-CRS has limited clinical utility due mainly to the lack of published psychometric data to support the tools usage. Although applicable to children and child friendly in its administration the broad range of movement disorders the scale attempts to cover may diminish its direct applicability specifically to children with dyskinetic cerebral palsy. A valuable addition to this scale is the General Assessment. The items in this section are developmentally appropriate for the designated age range and include information regarding attention and alertness during observation and at home, an area frequently influenced by pharmacologic intervention. The Movement Disorder Assessment that investigates severity is more complex to score and unlike all other dyskinesia assessments rates the presence of the movement disorder at rest as scoring only 1 (on the 0-4 scale), where all other scales score the presence of dyskinesia at rest as the most severe, i.e. a score of 4.

INTENDED POPULATION: Children and adolescents aged 4-18 years with movement disorders of primary and secondary aetiology including: Hypokinetic-rigid, Chorea/Ballism, Dystonia/Athetosis, Myoclonus, Tic, and tremor.

REPORTING STYLE: Observation and parent report.

ADMINISTRATION: A detailed video protocol (Table 6) taking approximately 20 minutes, is provided. In addition to short parental interview regarding alertness and attention, swallowing and drooling, self-feeding and personal care items is required.

Table 6: MD-CRS video protocol

PERSPECTIVE	ACTIVITY	TIME
Full body view	Child remove shoes, UL and LL garments	1 minute
	Supine position 1 minute	1 minute
	Sitting position (support trunk if required)	2 minutes
	Standing position (support if required) Walk 5m (with aids if required) Ask name or observe spontaneous speech Child put shoes, UL and LL garments back on	
Upper part of body (including ULs)	Sitting (chair, wc) in front of table Transfer five cubes L to R, R to L Draw person, write name	
Face: full body view	At rest or during activity Fix and follow visual stimulus Smile, open and close eyes Stick out tongue, move in all directions	

SCORING: The MD-CRS consists of two parts.

Part 1: General Assessment. Motor Function (head control, sitting, standing, walking, reaching, grasping and handwriting); Oral/Verbal Function (swallow, drooling and language); Self-Care (dressing, feeding and personal cares) and Attention/Alertness (alertness and attention during assessment and at home).

Part 2: Movement Disorder Severity. Assesses movement disorders across seven body regions (eyes and periorbital region, face, tongue and perioral region, neck, trunk, upper limbs and lower limbs). Both parts employ five-point ordinal scales (See Table 7 for scoring criteria).

Scores are calculated using statistical analysis to provide a Global Index measure (range 0–1) which is then assigned a Global Class. The 5 classes are: 0–0.2 healthy; 0.2–0.4 mildly affected; 0.4–0.6 moderately affected; 0.6–0.8 severely affected and 0.8–1 profoundly affected.

Table 7: Scoring criteria for MD-CRS

	SCORE	SCORING CRITERIA
Part 1: General Assessment	0	Normal
	1	Mildly affected by MD, occasional difficulties, minimal assistance
	2	Moderately affected by MD, partially dependent
	3	Severely affected by MD, fully dependent
	4	Absent, totally dependent
Part 2: Movement Disorder Severity	0	MD absent
	1	MD present only at rest
	2	MD present during one / some tasks for region and/or involves 1–2 other regions
	3	MD present during one / some tasks for region and/or involves > 3 regions
	4	MD present during all tasks for region and/or involves ≥ 3 other regions, completion impossible

6.5 MOVEMENT DISORDER – CHILDHOOD RATING SCALE 0-3 (MD-CRS 0-3)

DESCRIPTION: The MD-CRS 0-3[41] is a version of the MD-CRS developed for children under the age of four years. The scale consists of the same two parts as the MD-CRS but is directed more towards the typical developmental stages of younger children. Part 1, the General Assessment and consists of Motor Function, Oral/Verbal Function and Attention/Alertness. Part 2 assesses Movement Disorder Severity across seven body regions.

PSYCHOMETRIC PROPERTIES: No articles reporting on the validity or reliability of the MD-CRS (0-3) specifically in children with cerebral palsy are available. One medication trial study[69] indicates the tool is suitable to detect change following an intervention in children with cerebral palsy.

CLINICAL UTILITY: The MD-CRS (0-3) has limited clinical utility due mainly to the lack of published psychometric data to support its use. Although applicable to small children and child friendly in its administration the broad range of movement disorders the scale attempts to cover may diminish its direct applicability specifically to children with dyskinetic cerebral palsy. A valuable addition to the scale is the General Assessment. The items in these sections are developmentally appropriate for the young children.

INTENDED POPULATION: Children and adolescents aged 0-3 years with movement disorders of primary and secondary etiology including: Hypokinetic-rigid, Chorea/Ballism, Dystonia/Athetosis, Myoclonus, Tic, and tremor.

REPORTING STYLE: Observation and parent report.

ADMINISTRATION: A detailed video protocol is provided (Table 8). A short parental interview regarding alertness and attention, swallowing and drooling is also required.

Table 8: MD-CRS (0-3) video protocol

PERSPECTIVE	ACTIVITY	TIME
Full body view	Supine position	1 min
	Sitting position (support trunk if required)	1 min
	Standing position (support if required)	2 min
	Walk 3m (with aids if required)	
	At rest or during activity	
Upper part of body (including ULs)	Sitting (chair, mothers lap, wc) Present rattle, observe reach and grasp	
Face: full body view	At rest or during activity	

SCORING: The MD-CRS 0-3 consists of two sections:

Part 1: General Assessment. Motor Function (head control, sitting, standing, walking, reaching, grasping, Oral/Verbal Function (swallowing and drooling), and Attention/Alertness (during observation and at home). This is scored on a five-point ordinal scale.

Part 2: Movement Disorder Severity. Assesses movement disorders across seven body regions (eye and periorbital, face, tongue and perioral, neck, trunk, upper limbs and lower limbs) using a three-point ordinal scale: movement disorder is absent, intermittent or constant (see Table 9 for scoring criteria).

Scores are calculated using statistical analysis to provide a Global index measure (range 0-1) which is then assigned a Global Class. The five classes are: 0-0.2 healthy; 0.2–0.4 mildly affected; 0.4-0.6 moderately affected; 0.6-0.8 severely affected and 0.8-1 profoundly affected.

Table 9: Scoring criteria for MD-CRS(0-3)

	SCORE	SCORING CRITERIA
Part 1: General Assessment	0	Normal
	1	Mildly affected by MD, occasional difficulties, minimal assistance
	2	Moderately affected by MD, partially dependent
	3	Severely affected by MD, fully dependent
	4	Absent, totally dependent
Part 2: Movement Disorder Severity	0	MD absent
	1	MD is intermittent
	2	MD is constant

6.6 DYSKINESIA IMPAIRMENT SCALE (DIS)

DESCRIPTION: The DIS[42] is the most recently published scale. It was designed to measure and differentiate between dystonia and choreoathetosis at rest and on activity specifically in people with dyskinetic cerebral palsy. The DIS is the only scale to address both dystonia and choreoathetosis, movement disorders that frequently occur concurrently in children with dyskinetic cerebral palsy[16,27,44]. The DIS evaluates dystonia and/or choreoathetosis across 12 body regions, rating duration dystonia/choreoathetsosis is present and amplitude of the movement.

PSYCHOMETRIC PROPERTIES: Authors of the DIS have reported that it distinguishes and quantifies between dystonia and choreoathetosis but there are no independent studies investigating this capability. Moderate internal consistency and fair inter-rater reliability[42,70] are evident with no evidence of intra-rater, test-retest reliability or responsiveness to change. Limited content validity[42] and limited concurrent[42] and predictive validity are demonstrated[43]. The tool has not yet been used in a published outcome study and no responsiveness to change data is currently available.

CLINICAL UTILITY: The DIS is a lengthy and complex assessment tool to both video and score, limiting its overall clinical utility. The tool is available in the journal article with adequate instructions for scoring and a detailed video protocol[42]. The video protocol, takes a maximum of 30 minutes and then an additional 30 to 45 minutes per subscale is required for scoring with the addition of range of motion being assessed as this is required for scoring. Clinical experience with complex movement disorders in children with cerebral palsy is required for reliable use of this scale and its length lends itself to a comprehensive research tool rather than for use in clinical practice.

INTENDED POPULATION: People with dyskinetic cerebral palsy.

REPORTING STYLE: Observational and ROM measurement required to score.

ADMINISTRATION: A detailed video protocol accompanies the DIS (Table 10), from which the tool can be scored. Videoing takes a maximum of 30 minutes plus an additional 30 to 40 minutes to score. Passive range of motion via goniometry is required for assessment of amplitude.

Table 10: DIS video protocol

POSITION	ACTIVITY	VIDEO VIEW
General	Enter room (walk, wc)	Front
Sitting: comfort position	Sit at rest (chair or wc)	Front & close
	Eyes tracking movement	Close up
	Eye blinking x10	Close up
	Open and close mouth x10	Close up
	Speech	Bust
	Turn head R & L	Bust
	Latero-flexion head to R & L x5	Bust
	Elevate ULs sidewards x5	Front
	Grasp cup, move R to L of table, R & L hand	Front
	Grasp pen, move R to L of table, R & L hand	Front
Sitting: active position	Active sitting on bench	Front & profile
	Bend trunk forwards and back x5	Front & profile
Lying position	Lying on mat	Front
	Grasp/reach pen (R & L hand) side to overhead, cross midline	Front
	Roll right and left	Front
Standing position	Stand upright	Front, left & right profile
Each task 30 seconds, maximum 30 minutes		

SCORING: The DIS evaluates dystonia and/or choreoathetosis across 12 body regions: eyes (tracking, blinking); mouth (open/close, speech); neck (lateroflexion, rotation); trunk (active sitting, forward flexion); right and left proximal arm (abduction, grasp and move pen); right and left distal arm (abduction, grasp and move pen); right and left proximal leg (rolling, standing) and distal leg (rolling, heel/toe raising).

It rates on a five-point ordinal scale for duration, the percentage of time dystonia or choreoathetosis are present and amplitude of observed movement as percentage of range the dystonia or choreoathetosis occurs within (see Table 11 for scoring criteria). The total maximum score for each of the dystonia and choreoathetosis scales is 288, comprised of Duration and Amplitude scores on action (maximum of 192) and at rest (maximum score of 96).

Table 11: Scoring criteria for the DIS

	SCORE	SCORING CRITERIA
Duration Factor	0	D/CA is absent
	1	D/CA is occasionally present (<10%)
	2	D/CA is frequently present (≥10 – <50%)
	3	D/CA is mostly present (≥50 – <90%)
	4	D/CA is always present (≥90%)
Amplitude Factor	0	D/CA is absent
	1	D/CA in small ROM (<10%)
	2	D/CA in moderate ROM (≥10 – <50%)
	3	D/CA in submaximal ROM (≥50 – <90%)
	4	D/CA in maximal ROM (≥90%)

6.7 KINEMATIC DYSTONIA MEASURES

Kinematic analysis of dyskinetic movement is emerging as a useful tool to quantify the amount and type of movement present in the limbs of children with dyskinetic cerebral palsy. Kinematic dystonia measures have been developed for laboratory based research into upper limb assessment[60,61]. They measure dystonia as elicited by voluntary movement during motor tasks. Involuntary movements are measured via a motion analysis system capturing wrist flexion/extension, forearm pronation/supination, elbow flexion/extension, shoulder flexion/extension, abduction/adduction and internal/external rotation. These systems are based on the premise that involuntary movement and upper limb postures are a feature of dystonia and these increase as the individual engages in voluntary actions.

Gordon et al (2006)[60] utilised kinematic analysis to assess and quantify dystonia in children with cerebral palsy presenting with both spasticity and dystonia. Dystonia was elicited using finger tapping of the contralateral limb. The findings from Gordon et al study[60] from n=13 children with cerebral palsy aged 7-17 years, demonstrated significant correlation between the kinematic measure of dystonia and BADS scores ($r=0.75$, $p<0.005$). Reach was also analysed using kinematic analysis and it was found children with more dystonia made more curved reach paths.

The Kinematic Dystonia Measure[61] captures joint positions and angular rotations of the shoulder, elbow and wrist using a motion-capture system. Changes in position and angles are then summed to provide a Kinematic Dystonia Measure score. Movement and abnormal posturing of the dystonic upper limb is elicited by tapping of the less affected or unaffected hand to the beat of an auditory cue thus enabling quantification of involuntary movement triggered by voluntary movement. Where hand tapping was not possible due to the presence of mirror movements or due to significant motor impairment, eye blinking in time to an auditory cue was used to illicit involuntary movement. Kawamura et al (2012)[61] found in their study of n=11 children aged 4.3 to 15.4 years with spasticity and dystonia from various diagnoses, Kinematic Dystonia Measure scores correlated with total BADS scores ($r=0.79$, $p=0.003$) and with affected upper limb BADS scores ($r=0.76$, $p=0.007$).

Kinematic analysis demonstrates that spasticity and dystonia are able to be quantified separately in those children that present with mixed hypertonia. These measures are currently laboratory based and have not yet been applied to detect change pre and post an intervention targeting reduction in dystonia.

6.8 DYSTONIA AND MOVEMENT DISORDER SCALES: DIAGNOSTIC GROUPS OTHER THAN CEREBRAL PALSY

A number of different scales have been developed to assess different movement disorder presentations across multiple diagnostic groups. The majority of these provide impairment level information regarding the severity of the movement disorder with some also providing some insight into the impact of the movement disorder and/or disease on activities of daily living, behaviour and cognition as well as the assessment of pain due to the movement disorder. Although there is no evidence of their application to children with cerebral palsy in the literature, aspects of some of these scales could be applied to the assessment of dyskinesia in children with cerebral palsy. Scales that could provide some insight into aspects of movement disorder severity and assessment for children with cerebral palsy include: The Global Dystonia Scale[39]; The Unified Huntington's Disease Rating Scale[71]; Federal University of Minus Gerais Sydenham's Chorea Rating Scale[72]; Toronto Western Spasmodic Torticollis Rating Scale[73]; Brief Ataxia Rating Scale[74] and the Unified Myoclonus Rating Scale[75].

7. ASSESSMENT OF DYSKINETIC CEREBRAL PALSY ACROSS THE DOMAINS OF THE INTERNATIONAL CLASSIFICATION OF FUNCTIONING, DISABILITY & HEALTH

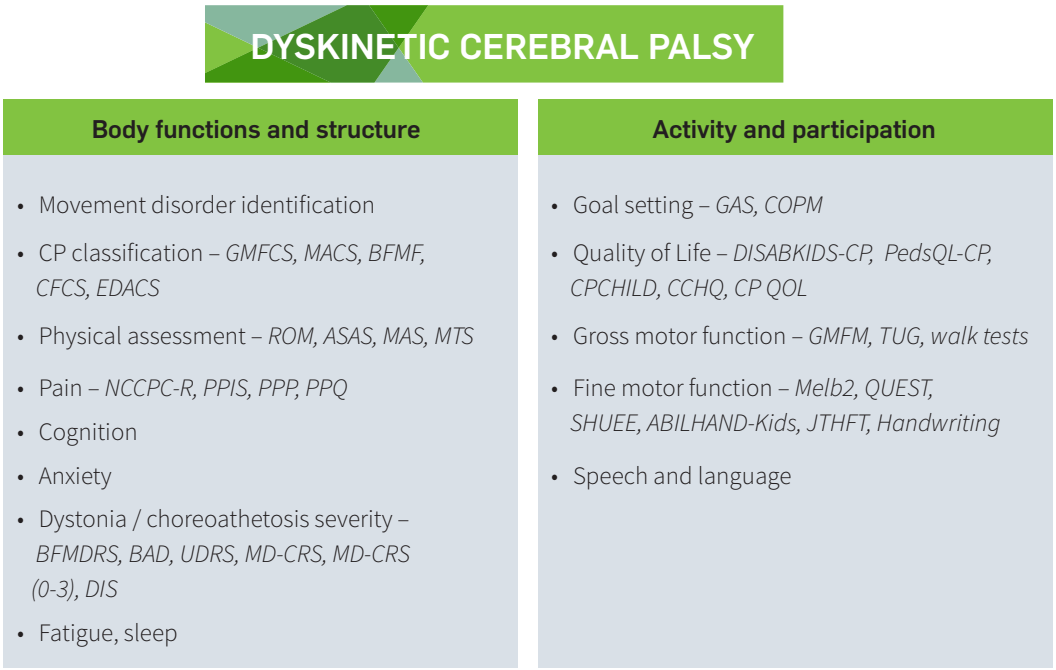
Dyskinesia scales provide impairment level outcome data with some scales providing select insight into the impact of the dyskinetic movement disorder on specific activities. Higher scores on all the scales indicate increasing severity of dyskinesia and reduced activity ability. Dyskinesia scales can be utilised for two purposes. First, they provide an indication of dyskinesia severity at a single time point and second, they can be utilised to measure responsiveness to change following a specific intervention aimed to target dystonia and or choreoathetosis. However, a reduction in dystonia and/or choreoathetosis does not necessarily translate to enhanced function or participation or improvements in caregiver burden and quality of life.

The International Classification of Functioning, Disability and Health (ICF)[4] provides clinicians with a framework to guide understanding of the complex interaction between the health condition, dyskinetic cerebral palsy, and the contextual factors of the environment as well as personal factors. This framework can also be utilised to guide assessment and ensure that intervention targets those areas most important to the person with dyskinesia.

Assessment of the child with cerebral palsy should be holistic and consider all domains of the ICF. It is important to assess not only body functions and structures but also activity and participation domains considering individual personal, cultural and environmental factors (Figure 4). Assessment of dyskinesia utilising one or more of the specifically designed scales should be complimented by other standardised assessments that provide information about quality of life, caregiver burden, independence in activities of daily living and individualised goals[43]. A study by Lumsden et al (2015)[76] found that few interventional studies utilised assessments or scales that objectively addressed the main concerns of children with dystonia and their carers. Monbaliu et al (2016)[43] found that dystonia has a greater negative impact on activity, participation and quality of life than choreoathetosis. A combined assessment approach provides greater detail than impairment level data alone on the impact of dyskinesia on the lives of children with cerebral palsy. 'The effectiveness of interventions should be judged using meaningful outcome measures that reflect the goals and expectations of individual patients'[77].

Additional assessment tools that could be utilised for children with dyskinetic cerebral palsy are included below. The selection of these tools was based on literature review and a survey of clinicians attending a symposium on the topic of Dyskinetic Cerebral Palsy organised through the Centre of Research Excellence in Cerebral Palsy (March 2017). A summary of the survey results can be found in Appendix 3. Assessment should also include questions around the areas of sleep patterns, levels of fatigue, bowel habits and pain as issues such as poor sleep, constipation, pain, anxiety and fatigue can exacerbate dyskinetic movements.

Figure 4: Assessment of dyskinetic cerebral palsy and the International Classification of Functioning, Health and Disability



7.1 PHYSICAL ASSESSMENT

Physical assessment of children with dyskinetic and mixed cerebral palsy should include assessment of range of motion, passive and active (if relevant) as dystonia and spasticity both affect range of motion which in turn impact function and participation. Some assessment of strength and selective motor control may also be indicated prior to specific interventions such as Botulinum toxin-A injections, intrathecal baclofen and medication trials which aim to decrease hypertonia. If hypertonia is decreased through medical interventions and underlying muscle strength is poor, function can be severely hampered in the more functional child with dyskinetic cerebral palsy.

SPASTICITY MEASUREMENT

Spasticity is one of the most commonly measured outcomes in people with cerebral palsy. Reliable and valid quantification of spasticity is important not only for clinical decision making and reliable evaluation of interventions but also for research database purposes. Methods reported in the literature for measuring spasticity include the Modified Tardieu Scale (MTS)[78], Modified Ashworth Scale (MAS)[79] and The Australian Spasticity Assessment Scale (ASAS)[80]. As spasticity frequently co-exists with dystonia and/or choreoathetosis in both dyskinetic and mixed cerebral palsy types, the assessment of spasticity is important, in combination with assessment of dystonia, as most interventions target both motor types.

The MTS quantifies the severity of muscle spasticity. It consists of two measures. The first, referred to as R2, is the maximum passive range of motion of the target muscle group. The second, R1, is elicited by moving the target muscle group from its shortest to longest position using a rapid velocity stretch. R1 is the angle at which muscle resistance or 'catch' is felt in response to the stretch. Both angles are measured with a goniometer. A catch early in the available range indicates more significant spasticity than a catch toward the end of the range. The relationship between R1 and R2 is important and indicates the dynamic component of spasticity.

The MAS measures the resistance of a muscle to passive movement. The muscle group is moved through its range of motion over a period of one second. Muscle response is graded on a six-point scale that describes the muscle resistance, the presence or absence of a catch, and the ease with which the joint is able to be moved through the available range. The MAS has documented limitations and poor reliability.

The ASAS is a relatively new clinical measure of spasticity for people with cerebral palsy, developed to provide unambiguous, tessellated criteria for scoring spasticity. Muscle groups are subjected to rapid passive stretch and a five-point scale grades the absence of a catch, or if a catch is present whether the catch occurs in the first or second half of the available range, as well as any resistance felt throughout the remaining range.

A reliability study indicates it is a promising tool to identify and quantify spasticity and to assist in classifying motor disorders for clinical, research, and epidemiological purposes[81].

Additional aspects of the physical assessment should also include height and weight. It is anecdotally noted that decreasing dystonia and/or choreoathetosis via medical interventions such as ITB, DBS and medications can impact weight gain due to a reduction in the amplitude, frequency and severity of movements.

7.2 INDIVIDUALISED GOAL ATTAINMENT

Individualised goal setting is generally acknowledged as an integral aspect of activity-focused interventions[82] and collaboratively developed, family centred goals focus the attention of those involved in the intervention on outcomes and improve motivation to participate in therapy[83]. Goal setting was unanimously selected as very important to consider as part of a well-rounded assessment for children with dyskinetic cerebral palsy (see Appendix 3 for survey results from 113 clinicians attending an education symposium on dyskinetic cerebral palsy). There are several goal setting approaches that can be utilised to measure intervention outcomes for children with cerebral palsy. The two most commonly used include:

- Canadian Occupational Performance Measure (COPM)[59]
- Goal Attainment Scaling (GAS)[84]

The COPM is the most widely used goal setting instrument with sound psychometric evidence[85]. It is an individualised measure of a client's self-perception of their occupational performance and provides a structure to identify goals in the areas of self-care, productivity and leisure and measure outcomes based on individual performance and satisfaction with performance[59]. Goals are identified and the five most important occupational performance goals are rated on a ten-point scale for: Performance, how well they feel they can complete the activity; and Satisfaction with Performance, how satisfied they are with their current ability to complete the activity.

GAS is a methodology for tailoring intervention goals and measuring progress towards goal achievement. It can be used by itself or in combination with other assessments, such as COPM. Goals are determined in combination with therapists and families and scaled on a five-point scale. The scale generally rates baseline (or current) performance at (-2) and the desired or expected goal level following intervention at (0). A less than expected outcome is given a score of (-1) indicating progress towards goal attainment and then two additional levels of achievement, which exceed the expected outcome, are greater than expected (+1) and much greater than expected (+2).

7.3 QUALITY OF LIFE AND CAREGIVER BURDEN

Quality of life is a general concept and can be defined as 'an individuals' perception of their position in life, in the context of culture and value systems in which they live and in relation to their goals, expectations, standards and concerns'[86]. With cerebral palsy constituting the most common cause of chronic childhood disability, quality of life is an important construct to consider for all children with cerebral palsy as there is likely to be some impact not only on the physical but also the social and emotional wellbeing of the child and their family. Assessment of quality of life (QOL), health related quality of life (HQOL) and caregiver burden are complimentary to impairment and functional assessment of the child with dyskinetic cerebral palsy.

There are a number of QOL scales/questionnaires that have been developed to specifically measure QOL in children with cerebral palsy (Table 12). These include:

- DISABKIDS CP Module[87]
- Pediatric Quality of Life Inventory CP Module (PedsQL 3.0 CP)[88]
- Caregiver Priorities and Child Health Index of Disabilities (CPCHILD)[89]
- Care and Comfort Hypertonia Questionnaire (CCHQ)[90]
- Cerebral Palsy Quality of Life-Child (CP QOL-Child)[91]
- Cerebral Palsy Quality of Life-Child (CP QOL-Teen)[92]

Selection of the most appropriate QOL measure will frequently be guided by the functional capabilities of the child being assessed and the outcomes of interest, i.e. QOL, HQOL and/or caregiver burden. A systematic review of cerebral palsy condition specific QOL measures found the CP QOL and the CPCHILD had the strongest psychometric properties and clinical utility[93]. This review found the DISABKIDS and PedsQL 3.0 CP were moderately constructed with weaker psychometrics, while the CCHQ scored poorly as an outcome measure on the quality scale used[93]. A survey of clinicians regarding preferred QOL tools for children with dyskinetic cerebral palsy showed preference for the CPCHILD and CPQOL (Appendix 3). Recently presented responsiveness to change data on the CPCHILD pre and post orthopaedic surgery has strengthened its use as a sensitive outcome measure for children with more severe presentations of cerebral palsy[77].

The CPCHILD evaluates function and health status, caregiver burden and health related quality of life in children with severe cerebral palsy. It has been validated for use for caregivers of children with severe developmental disabilities such as those with non-ambulatory cerebral palsy and traumatic brain injury, who would be categorised in level IV or V of the GMFCS[89]. Responsiveness to change has been demonstrated following hip surgery in children with cerebral palsy, GMFCS levels IV and V[77] and spinal surgery for children with severe cerebral palsy[94]. The domains of the CPCHILD include: Personal Care, Positioning, Transferring and Mobility, Comfort and Emotions, Communication and Social Interactions and Health. It also comments on pain and the importance of QOL items to the child.

The CP QOL–Child is a cerebral palsy-specific questionnaire designed to be used for children between the ages of 4 and 12 years. The CP QOL–Teen was designed for children with cerebral palsy aged 13 to 18 years. The questionnaires have self-report and caregiver proxy versions. The CP QOL measures seven areas of a child's life: Social Wellbeing and Acceptance, Participation and Physical Health, Emotional Wellbeing, Pain and Impact of Disability, Access to Services and Family Health.

The CCHQ questionnaire was developed to evaluate the functional care needs, and to a lesser extent quality of life in children with increased tone of cerebral origin, particularly those with 'severe' cerebral palsy.

Early work on the CCHQ has been undertaken to establish content validity and the CCHQ has also been shown to be sensitive enough to detect changes when ITB was offered or dose levels changed. Formal evaluation of reliability and validity has not been finalised. It is a self-report questionnaire and requires parents or caregivers to rate how easy or difficult it is for them or their child, in the last two weeks, to perform a range of tasks relative to a cooperative person without a disability. The domains covered include: Personal Care, Positioning/Transferring and Comfort and Interaction/Communication.

The DISABKIDS CP Module is a condition specific module of the DISABKIDS project. It focuses on the impact of cerebral palsy on QOL with two additional questions around communication. The DISABKIDS chronic generic module was designed to assess QOL for any child, aged 4–16 years, with a chronic health condition and can be used in addition to the cerebral palsy module. The scale provides score between 1–100, with higher scores indicating better quality of life and better adjustment to cerebral palsy.

The PedsQL 3.0 CP Module is a cerebral palsy-specific QOL instrument designed for children 2–18 years with child self-report and parent proxy report formats. The questionnaire consists of 35 questions across seven domains: Daily Activities, School Activities, Movement and Balance, Pain and Hurt, Fatigue, Eating Activities, Speech and Communication. Not all items are applicable to the younger ages. Responses are scored on a Likert scale (0 = never a problem; 1 = almost never a problem; 2 = sometimes a problem; 3 = often a problem; 4 = almost always a problem) with higher scores indicating better HRQOL, fewer symptoms or problems.

Table 12: **Overview of QOL measures for children with cerebral palsy**

TOOL	POPULATION	ASSESSES	CLINICAL UTILITY / PSYCHOMETRICS
CPCHILD	Severe CP	Personal Care, Positioning, Transferring and Mobility, Comfort and Emotions, Communication and Social Interactions and Health	Strong
CPQOL-Child CPQOL-Teen	CP specific	Social Wellbeing and Acceptance, Participation and Physical Health, Emotional Wellbeing, Pain and Impact of Disability, Access to Services, Family Health	Strong
CCHQ	CP	Personal Care, Positioning/Transferring, Comfort, Interaction/Communication	Weak
DISABKIDS CP Module	CP module 4 – 16 yrs	Impact of cerebral palsy on QOL and Communication	Moderate
PedsQL 3.0 CP	CP module 2 – 18 years	Daily, School and Eating activities, Movement and Balance, Pain and Hurt, Fatigue, Communication	Moderate

7.4 PAIN

Pain in children with cerebral palsy is under recognised and appears to be highly correlated with increasing GMFCS levels[95]. A study by Penner et al (2013)[96] found hip subluxation/dislocation and dystonia to be the most common causes of pain in children and youth with cerebral palsy. A systematic review on cerebral palsy[97] found that three in every four children with cerebral palsy experience pain, regardless of the level of their disability. The review also found pain is linked to lower participation levels and higher rates of behavioural problems and increases with age. Penner et al (2013)[96] investigated the characteristics of pain in children and youth with cerebral palsy and found more than 54% reported pain and that nearly 25% had pain that prevented some to most activities. A study into childhood dystonia by Lumsden et al (2015)[76] found pain to be the most commonly expressed concern for children and their carers. Pain, although not well reported and rarely measured in intervention studies in childhood dystonia[76] is thought to be a driver for dystonic movements, further impacting musculoskeletal pain in children with cerebral palsy.

The three approaches to pain measurement include self-report, observational and behavioural and physiologic. A systematic review of chronic pain assessment tools for cerebral palsy by Kingsnorth et al (2015)[98] found seven paediatric chronic pain tools that can be utilised to determine if a child has chronic pain, four having been used to specifically assess pain in children with cerebral palsy. The authors concluded that no one tool meets the needs of all children with cerebral palsy experiencing chronic pain. In a survey of clinicians regarding the importance of assessment of pain in children with dyskinetic cerebral palsy, it was deemed very important by most respondents (Appendix 3). All four tools specifically developed or adapted to assess pain in cerebral palsy were seen as offering value for children with dyskinetic cerebral palsy (Table 13).

These include:

- Noncommunicating Children's Pain Checklist-Revised (NCCPC-R)[99]
- Pediatric Pain Interference Scale (PPIS)[100]
- Pediatric Pain Profile (PPP)[101]
- Pediatric Pain Questionnaire (PPQ)[102]

The NCCPC-R can be utilised to assess past chronic pain, current pain and postoperative pain and has been validated with care givers of children with cerebral palsy and other cognitive and motor impairments. The tool requires two hours of observation and a 30 item questionnaire. It has strong psychometric properties and good clinical utility, although the two-hour observation period may limit its clinical use. It can be used for children across all levels of the GMFCS.

The PPIS, initially developed for adults to assess pain related behaviours across the domains of pain, fatigue, physical functioning, social health, and emotional health has now been validated for paediatric chronic health conditions (5–18 years) including cerebral palsy. It can be self report or parent proxy and has moderate clinical utility. It is most appropriate for GMFCS levels I to III as items relate to mobility and weight bearing.

The PPP was specifically developed to assess pain in children with neurologic impairments, including cerebral palsy. It is a 20-item evaluative and discriminative scale with well-established psychometric properties and moderate clinical utility. A systematic review found it did not fully capture the impact of pain on quality of life and function[98]. It is appropriate for all levels of the GMFCS.

The PPQ was developed to assess chronic pain from the perspective of the child and parent. It has been adapted to use with children with cerebral palsy. The presence and quality of a child's pain is indicated by a parents use of verbal and non-verbal cues. It has well established psychometric properties and moderate clinical utility but requires greater validation with GMFCS levels IV and V.

Table 13: Overview of pain tools for children with cerebral palsy

PAIN TOOL	POPULATION	ASSESSES	FORMAT	PSYCHOMETRICS	CLINICAL UTILITY
NCCPC-R	Validated for CP (GMFCS I-V)	Past, current and post-op pain	2-hour obs and 30 item questionnaire	Well established	Strong (2 hour obs may limit use)
PPIS (PROMIS)	Validated for chronic paed conditions incl CP (5-18yrs) GMFCS I-III	Pain behaviours: fatigue, physical, social and emotional health	Self report, parent proxy	Approaching well established	Moderate
PPP	Neurological impairments incl CP (GMFCS I-V)	Pain behaviours, past and current pain problems	20-item parent proxy scale	Well established	Moderate
PPQ	Adapted for CP (GMFCS I-III)	Chronic pain – intensity, quality and location	VAS for pain intensity and body diagram	Well established	Moderate

In addition to these pain tools self-report visual analogue scales such as the Wong-Baker Faces Pain Scale[103] (http://wongbakerfaces.org) can be useful in the clinical setting and are the preferred pain reporting tool for many clinicians (Appendix 3).

7.5 ACTIVITY AND PARTICIPATION

The range of assessments for children with cerebral palsy that assess aspects of activity and participation is extensive and choice is dependent upon the activity itself as well as the functional abilities of the child. For example, assessments can be used to measure: speed and quality of gait, gross motor function, fine motor function, independence in activities of daily living, developmental milestones, sleep, fatigue, speech and communication. Listed below are a selection of assessments that a survey of clinicians with a clinical interest in dyskinetic cerebral palsy selected for use specifically for children with dystonia and dyskinetic cerebral palsy (Appendix 3).

7.5.1 GROSS MOTOR AND MOBILITY ASSESSMENTS

Gross Motor Function Measure (GMFM)[58]

A criterion-referenced clinical measure designed to evaluate change in gross motor function in children with cerebral palsy. It has been proven to be reliable, valid and responsive to change. It assesses gross motor function in five dimensions: lying and rolling, sitting, crawling and kneeling, standing, walking, running and jumping. The two formats include the GMFM-88, reported to take 45-60 minutes and the GMFM-66, which takes 30-40 minutes.

Walk tests

Walk tests measure the walking capacity of a child over a set distance. They are easy, repeatable and objective measures. The 10-metre walk measures speed, stride length and cadence and the six-minute walk measures endurance. There are also one-minute and two-minute walk tests. The six-minute walk has been shown to have good reliability and sensitivity to change in adults with cerebral palsy[104] and good test-retest reliability in children with cerebral palsy[105,106].

Timed Up and Go (TUG)[107]

The TUG times how long it takes a child to stand from a seated position, walk three metres, return and sit down without physical assistance. It has good reliability for children with cerebral palsy[108].

Gillette Mobility Scale[109]

The Gillette Mobility Scale is a ten-level, parent-report walking scale assessing walking abilities from non-ambulatory to ambulatory in all community settings and terrains. It has established psychometric evidence and can be useful in assisting clinicians to document functional change in children with chronic neuromuscular conditions.

Additional gross motor assessments that could be considered include:

- The Quality FM1[10]: an observational assessment of Stand and Walk/Run/Jump skill items from the GMFM-66 and was designed for children with cerebral palsy aged five and over.
- The Challenge Module[111]: a 25-item assessment of advanced motor skills for children with cerebral palsy, GMFCS I and II over the age of six years.
- HiMAT[112]: a 13-item, high level mobility and balance assessment developed for adolescents and adults with acquired brain injury.

- Pediatric Balance Scale (PBS)[113], a modification of the Berg Balance Scale[114]: assesses functional balance skills in school-aged children with mild to moderate motor impairment.

The use of video for gait and mobility analysis, either formally or informally should always be considered in those children with independent mobility.

7.5.2 FINE MOTOR AND UPPER LIMB ASSESSMENTS

Quality of Upper Extremity Skills Test (QUEST)[115]

The QUEST is a reliable and valid measure for evaluating the quality of upper limb movement in children with cerebral palsy aged 18 months to 8 years. Domains include: dissociated movement, grasp, protective extension and weight bearing. The assessment focuses on patterns of movement that form the basis of developmental upper limb performance and is completed during a 30-45 minute structured session in the form of play. The QUEST is a reliable and valid measure for evaluating quality of movement in children with cerebral palsy.

Assisting Hand Assessment (AHA)[116] and Mini Assisting Hand Assessment (Mini AHA)[117]

The AHA, for ages 18 months to 12 years and the Mini AHA, for ages 8 months to 18 months, are criterion referenced outcome measures designed for use with hemiplegic children with cerebral palsy. They describe how effectively a child uses their hemiplegic or affected hand in collaboration with their non-affected hand during bimanual play. Assessment occurs during a video recorded 15-minute semi structured play session from which scoring is completed. Evidence of good validity, reliability and sensitivity to change is reported in various research studies.

Melbourne Assessment 2: Test of Unilateral Upper Limb Function (MA2)[118,119]

The MA2 is test of unilateral upper limb function that evaluates the quality of upper limb movement in children aged 2½ to 15 years with a neurological condition. It measures: movement range, accuracy, dexterity and fluency with 14 test items of reaching to, grasping, releasing and manipulating simple objects. It takes approximately 30 minutes to administer and 30 minutes to score from video. It has well established psychometrics.

If a questionnaire would be more appropriate, then the Children's Hand-Use Evaluation Questionnaire (CHEQ)[120] or the ABILHAND-Kids[121] may be useful[122]. The CHEQ is an online questionnaire developed for children with unilateral functional limitations. It evaluates and describes the experience of children in using their affected hand in bilateral activities. The ABILHAND-Kids is a parent questionnaire that measures the bimanual ability of children with cerebral palsy and their ability to manage daily activities.

7.5.3 SPEECH, LANGUAGE, COMMUNICATION AND ORAL MOTOR ASSESSMENT

The assessment of speech and communication in children with dyskinetic cerebral palsy will be dependent on oral motor ability and other comorbidities such as intellectual functioning. Speech production is frequently hampered or prevented by the presence of dystonia and the capacity of the child to engage in conversation should not be taken as an indication of their intellectual functioning. Concerns should be referred to a speech pathologist experienced with working with children with cerebral palsy for thorough assessment. This assessment may include alternative and augmentative communication as well as investigation of eating and drinking. The Viking Speech Scale[123] is one tool that can be used to classify speech production, alternative tools would be required for assessment of communication via alternative methods.

Viking Speech Scale: Classifies usual speech production in children with cerebral palsy aged over 4 years of age. It has been found to be a reliable measure of speech performance[124]. The scale has four levels:

Level I = not affected by motor disorder

Level II = speech imprecise but understandable to unfamiliar listeners

Level III = speech unclear, not understandable to unfamiliar listeners out of context

Level IV = no understandable speech

7.5.4 OTHER ASSESSMENTS RELEVANT TO CHILDREN WITH DYSKINETIC CEREBRAL PALSY

Depending on the child and their individual issues the assessment of independence in activities of daily living using tools such as the Pediatric Evaluation of Disability Inventory (PEDI)[57] and Pediatric Evaluation of Disability Inventory - Computer Adaptive Tests (PEDI-CAT)[125] or the Functional Independence Measure for Children (WeeFIMTM)(**https://www.udsmr.org/WebModules/WeeFIM/Wee_About.aspx**) may be appropriate. If saliva control is a problem a measure such as the Drooling Impact Scale[126] may be useful to determine both the extent of the problem and measure treatment effect. Fatigue is an ongoing issue for children with dyskinetic movements and dystonia due to their continuous and often extreme movement patterns. The assessment of the impact of fatigue on daily routines may be important. The Fatigue Assessment, one of the multidimensional scales from the PedsQLTM (**http://www.pedsql.org**), can be utilised to assess impact of fatigue. Other assessments could be utilised to measure anxiety, fatigue and sleep.

The PEDI assesses how a child functions with an impairment in the context of their daily life. It assesses functional skills, caregiver assistance and modifications required for self-care, mobility and social function. The PEDI-CAT is an online computerised version of the PEDI.

The WeeFIM was developed for children aged 6 months to 7 years with an acquired or congenital disease as an indicator of disability severity and to track change during rehabilitation. It comprises a checklist that rates the amount of assistance required for performance of activities in the areas of self-care, mobility and cognition. It is a valid and reliable assessment tool.

The Drooling Impact Scale evaluates the impact of drooling in children with developmental disabilities and is sensitive to changes in drooling in response to saliva control interventions. The questionnaire can be completed by a carer or via interview. The scale is valid, reliable and responsive to change.

Fatigue Assessment is one of the Multidimensional scales from the PedsQLTM. It investigates: General Fatigue, Sleep/Rest Fatigue and Cognitive Fatigue.

8. INTERVENTIONS FOR CHILDREN WITH DYSKINETIC CEREBRAL PALSY

Children with dyskinetic cerebral palsy tend to have more severe motor impairment compared to those children with spastic cerebral palsy. A prevalence and severity study from the SCPE[16] found almost 60% required a wheelchair and more than half had co-morbidities such as severe learning disability and epilepsy. A study by Monbaliu et al (2017)[127] that profiled the functional classification levels of a cohort of children with dyskinetic cerebral palsy found 79% were classified at GMFCS levels IV and V, 77% at MACS levels IV and V. This was in contrast to only 20% being classified at CFCS levels IV and V. The usually combined presence of spasticity and dystonia/choreoathetosis in children with dyskinetic cerebral palsy makes treatment and medical interventions more challenging[128]. Treatment and prognosis for dystonia, compared to spasticity, differ[129] and it is generally recognised that there is no established gold standard for the treatment of dyskinetic cerebral palsy[130].

Interventions commonly used for spastic cerebral palsy are often anecdotally deemed unpredictable in children with dystonia[128] and few papers address any interventions specifically aimed to address choreoathetosis. There are few interventions aimed specifically towards children with dyskinetic cerebral palsy, although many medical interventions do target dystonia reduction in addition to spasticity management. These include Intrathecal Baclofen (ITB), Botulinum toxin-A injections (BoNT-A), pharmacological interventions and surgery. Deep Brain Stimulation (DBS) specifically targets dystonia with well documented results in those of primary dystonia aetiology but less well supported results in secondary dystonias.

With a lack of disease modifying therapies to target dystonia and dyskinetic cerebral palsy, treatment goals need to target symptomatic relief of abnormal movement postures, associated pain and discomfort and other comorbidities such as orthopaedic complications, depression and anxiety. Treatment selection is generally based on severity and distribution and is based on empirical observation and experience as robust randomised controlled trials in this population are lacking.

8.1 THERAPY

The role of physical therapies in dyskinetic cerebral palsy, including occupational therapy and physiotherapy, is generally to maintain passive range of motion, minimise contractures, maximise function and ease burden of cares. Occupational therapists, speech pathologists and physiotherapists explore environmental modifications to ease caregiver burden and/or maximise independence in the areas of mobility, hand function, speech and communication and activities of daily living. This may involve the trial, prescription and modification of equipment in addition to child and family education around tips and tricks for living with dystonia.

There is currently no evidence base for interventions specifically aimed at children with dyskinetic cerebral palsy although the evidence for interventions for children with cerebral palsy should be considered[131]. Therapy interventions for children with dyskinetic cerebral palsy may include, but are not limited to:

- Hippotherapy — to work on improving trunk stability and symmetry.
- Hydrotherapy — to work on motor control and relaxation.
- Assistive technology — for communication, education, leisure and environmental control. Good positioning and alternative access may need to be investigated, including eye gaze technology.
- Seating and positioning — seating and positioning systems offering good proximal stability can enhance distal control. Alternative position systems are frequently required to enable change of position such as standing frames, sleep systems and alternative positioning chairs.
- Orthoses and casting — are frequently not well tolerated in children with dyskinetic cerebral palsy. Additional padding may be required and the purpose of the orthoses or cast carefully considered.
- Dysphagia management.
- Education — for both the child and family around triggers that can exacerbate movements, such as pain, stress, constipation, lack of sleep and managing these.

8.2 BOTULINUM TOXIN-A INJECTIONS (BONT-A)

Botulinum toxin-A is a neurotoxin injected into targeted muscles to treat localised spasticity and dystonia in children with cerebral palsy. BoNT-A blocks the release of acetylcholine, one of the main neurotransmitters at the neuromuscular junction and causes muscle paralysis. This paralysis, or muscle weakness usually lasts between three and six months, when repeat injections may be indicated.

BoNT-A injections are considered following careful functional and/or carer goal identification and the appropriate dystonic and/or spastic muscles are targeted. Current literature indicates there is strong evidence to support the use of BoNT-A injections for upper and lower limb spasticity management in cerebral palsy. There is still insufficient evidence to support its use in improving motor function[131,132]. There is evidence to support the use of BoNT-A for focal dystonia[133] but little evidence to specifically support its use with dyskinetic cerebral palsy.

8.3 INTRATHECAL BACLOFEN (ITB)

Baclofen is a commonly trialled oral medication for children with generalised dystonia and spasticity. Its action on receptors in the spinal cord suppresses muscle spasms and reduces muscle tone. In oral form, it crosses the blood brain barrier poorly which can necessitate higher doses producing unwanted side effects. Administered intrathecally, baclofen can be delivered directly to the site of action, allowing smaller doses and fewer side effects. An ITB pump, consisting of a programmable pump and intrathecal catheter, can administer a continuous infusion plus or minus bolus doses of baclofen[66,134]. ITB has significant complication rates with between 20 to 30% of cases experiencing adverse events or complications[135] and children with dystonia predominant presentations experiencing significantly higher complication rates than those children with spasticity predominant presentations of cerebral palsy[65].

There is currently weak evidence to support the administration of ITB to help with the reduction of spasticity and dystonia in cerebral palsy[136]. Weak evidence also exists to support its use in improving health related quality of life outcomes[131,136].

8.4 SURGERY

Orthopaedic interventions are frequently used to treat musculoskeletal deformity in children with cerebral palsy. Children with dyskinetic cerebral palsy are at risk of developing secondary musculoskeletal deformities that impact mobility and joint congruity. Historically, orthopaedic intervention for children with cerebral palsy and dystonia has been deemed unpredictable, harmful and results in poor outcomes[128]. A study by Blumetti et al (2017)[128] of n=37 children with dystonic cerebral palsy, representative across the GMFCS, demonstrated that the results of lower limb orthopaedic surgery specifically in children with dystonia are not as unpredictable as historically and anecdotally believed and functional mobility and hip morphology can be improved.

8.5 MEDICATIONS

A variety of oral medications are routinely prescribed for children with cerebral palsy for the generalised reduction in spasticity and/or dystonia. Pharmacological treatments can be unsatisfactory and side effects frequently limit dose. Medications prescribed for the treatment of generalised dystonia include: Baclofen, Haloperidol, Levodopa, Tetrabenazine and Benzhexol. Many of these medications can have side effects such as drowsiness, sedation and weakness. Limited evidence is available to support or refute the use of medications for the treatment of generalised dystonia in cerebral palsy.

8.6 DEEP BRAIN STIMULATION (DBS)

DBS is a neurosurgical technique involving the implantation of electrodes into specific areas of the brain, usually the Globus pallidus and/or subthalamic nucleus. The electrodes are attached to a neuro-stimulator, implanted below the patient's clavicle(s) or in their lower abdomen. DBS aims to decrease dyskinetic/dystonic movements and improve health related quality of life. DBS has been effectively used in the treatment of pain since the 1960s and in the control of primary dystonias common in Parkinson's disease[137]. DBS has been used, with varying results, in patients with cerebral palsy and secondary dystonias over the past decade[137]. Currently there is limited evidence and only a small number of studies available to support this intervention, although DBS has been shown to be an effective treatment option for dyskinetic cerebral palsy[137].

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10. APPENDIX 1

RATING SCALES FOR CHILDREN WITH DYSKINETIC CEREBRAL PALSY

Rating scales, developed for use with patients with primary and secondary dystonia and/or choreoathetosis that have been utilised specifically with children with cerebral palsy:

- Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS)
- Barry Albright Dystonia Scale (BADS)
- Unified Dystonia Rating Scale (UDRS)
- Movement Disorder – Childhood Rating Scale (MD-CRS)
- Movement Disorder – Childhood Rating Scale 0-3 (MD-CRS 0-3)
- Dyskinesia Impairment Scale (DIS)

THE BURKE-FAHN-MARSDEN DYSTONIA RATING SCALE (BFMDRS)

Burke RE, Fahn S, Marsden CD, Bressman SB, Moskowitz C & Friedman J. (1985)

REGION	PROVOKING FACTOR	SEVERITY FACTOR	WEIGHT	SCORE
Eyes	0 No dystonia at rest or with action 1 Dystonia only with particular action 2 Dystonia with many actions 3 Dystonia on action of distant part of body or intermittently at rest 4 Dystonia present at rest	0 No dystonia 1 Slight: Occasional blinking 2 Mild: Frequent blinking without prolonged spasms of eye closure 3 Moderate: Prolonged spasms of eyelid closure, but eyes open most of the time 4 Severe: Prolonged spasms of eyelid closure, with eyes closed at least 30% of time	0.5	
Mouth	0 No dystonia at rest or with action 1 Dystonia only with particular action 2 Dystonia with many actions 3 Dystonia on action of distant part of body or intermittently at rest 4 Dystonia present at rest	0 No dystonia present 1 Slight: Occasional grimacing or other mouth movements (e.g. jaw opening or clenched, tongue movement) 2 Mild: Movement present less than 50% of time 3 Moderate dystonic movements or contractions present most of the time 4 Severe dystonic movements or contractions present most of the time	0.5	
Speech & Swallow	0 No dystonia at rest or with action 1 Occasional, either or both 2 Frequent, either 3 Frequent one and occasional other 4 Frequent both	0 No dystonia 1 Slightly involved: speech easily understood or occasional choking 2 Some difficulty in understanding speech or frequent choking 3 Marked difficulty in understanding speech or inability to swallow firm foods 4 Complete/almost complete anarthria or marked difficulty in swallowing foods or liquids	1.0	
Neck	0 No dystonia at rest or with action 1 Dystonia only with particular action 2 Dystonia with many actions 3 Dystonia on action of distant part of body or intermittently at rest 4 Dystonia present at rest	0 No dystonia 1 Slight: Occasional pulling 2 Obvious torticollis but mild 3 Moderate pulling 4 Severe pulling	0.5	
Right arm	0 No dystonia at rest or with action 1 Dystonia only with particular action 2 Dystonia with many actions 3 Dystonia on action of distant part of body or intermittently at rest 4 Dystonia present at rest	0 No dystonia present 1 Slight: clinically insignificant 2 Mild: obvious dystonia but not disabling 3 Moderate: able to grasp, with some manual function 4 Severe: no useful grasp	1.0	
Left arm	0 No dystonia at rest or with action 1 Dystonia only with particular action 2 Dystonia with many actions 3 Dystonia on action of distant part of body or intermittently at rest 4 Dystonia present at rest	0 No dystonia present 1 Slight: clinically insignificant 2 Mild: obvious dystonia but not disabling 3 Moderate: able to grasp, with some manual function 4 Severe: no useful grasp	1.0	
Right leg	0 No dystonia at rest or with action 1 Dystonia only with particular action 2 Dystonia with many actions 3 Dystonia on action of distant part of body or intermittently at rest 4 Dystonia present at rest	0 No dystonia present 1 Slight: but not causing impairment, clinically insignificant 2 Mild: walks briskly and unaided 3 Moderate: severely impairs walking and requires assistance 4 Severe: unable to stand or walk on involved leg	1.0	
Left leg	0 No dystonia at rest or with action 1 Dystonia only with particular action 2 Dystonia with many actions 3 Dystonia on action of distant part of body or intermittently at rest 4 Dystonia present at rest	0 No dystonia present 1 Slight: but not causing impairment, clinically insignificant 2 Mild: walks briskly and unaided 3 Moderate: severely impairs walking and requires assistance 4 Severe: unable to stand or walk on involved leg	1.0	
Trunk	0 No dystonia at rest or with action 1 Dystonia only with particular action 2 Dystonia with many actions 3 Dystonia on action of distant part of body or intermittently at rest 4 Dystonia present at rest	0 No dystonia present 1 Slight bending, clinically insignificant 2 Definite bending, but not interfering with standing or walking 3 Moderate bending, interfering with standing or walking 4 Extreme bending of trunk preventing standing or walking	1.0	

REGION	PROVOKING FACTOR	SEVERITY	WEIGHT FACTOR	PRODUCT	
Eyes	0 - 4	X	0 – 4	0.5	0 – 8
Mouth	0 - 4	X	0 – 4	0.5	0 – 8
Speech & Swallow	0 - 4	X	0 – 4	1.0	0 – 16
Neck	0 - 4	X	0 – 4	0.5	0 – 8
Right arm	0 - 4	X	0 – 4	1.0	0 – 16
Left arm	0 - 4	X	0 – 4	1.0	0 – 16
Right leg	0 - 4	X	0 – 4	1.0	0 – 16
Left leg	0 - 4	X	0 – 4	1.0	0 – 16
Trunk	0 - 4	X	0 – 4	1.0	0 – 16
				Sum:	
				Max = 120	

DISABILITY SCALE	
A: Speech 0 – Normal 1 – Slightly involved, easily understood 2 – Some difficulty in understanding 3 – Marked difficulty in understanding 4 – Complete/almost complete anarthria	**E: Hygiene** 0 – Normal 1 – Clumsy, independent 2 – Needs help with some activities 3 – Needs help with most activities 4 – Needs help with all activities
B: Handwriting 0 – Normal 1 – Slight difficulty, legible 2 – Almost legible 3 – Illegible 4 – Unable to grasp to maintain hold on pen	**F: Dressing** 0 – Normal 1 – Clumsy, independent 2 – Needs help with some activities 3 – Needs help with most activities 4 – Needs help with all activities
C: Feeding 0 – Normal 1 – Uses tricks, independent 2 – Can feed but not cut 3 – Finger food only 4 – Completely dependent	**G: Walking** 0 – Normal 1 – Slightly abnormal, hardly noticeable 2 – Moderately abnormal, obvious to naive observer 3 – Considerably abnormal 4 – Needs assistance to walk 6 – Wheelchair bound
D: Eating/swallowing 0 – Normal 1 – Occasional choking 2 – Chokes frequently, difficulty swallowing 3 – Unable to swallow firm foods 4 – Marked difficulty swallowing soft food and liquid	**Disability Scale Score:** ..... (Max = 30)

BARRY ALBRIGHT DYSTONIA SCALE (BADS)

Barry MJ, Van Swearingen JM & Albright AL. (1996)

EYES: Signs of dystonia in the eyes include prolonged eyelid spasms and/or forced eye deviations

0 – Absence of eye dystonia

1 – Slight: Dystonia less than 10% of the time and does not interfere with tracking

2 – Mild: Frequent blinking without prolonged spasms of eye closure and/or movements less than 50% of the time

3 – Moderate: Prolonged spasms of eyelid closure, but eyes open most of the time and/or eye movements more than 50% of the time that interfere with tracking but able to resume tracking

4 – Severe: Prolonged spasms of eyelid closure, with eyes closed at least 30% of the time and/or eye movements more than 50% of the time that prevent tracking

* Unable to assess eye movements

EYES: .....

MOUTH: Signs of dystonia in the mouth include grimacing of the mouth

0 – Absence of mouth dystonia

1 – Slight: Dystonia less than 10% of the time and does not interfere with speech and/or feeding

2 – Mild: Dystonia less than 50% of the time and does not interfere with speech and/or feeding

3 – Moderate: Dystonia more than 50% of the time and/or dystonia that interferes with speech and/or feeding

4 – Severe: Dystonia more than 50% of the time and/or dystonia that prevents speech and/or feeding

* Unable to assess mouth movements

MOUTH:.....

NECK: Signs of dystonia in the neck include pulling of the neck into any plane of motion: extension, flexion, lateral flexion or rotation

0 – Absence of neck dystonia

1 – Slight: Pulling less than 10% of the time and does not interfere with lying, sitting, standing and/or walking

2 – Mild: Dystonia less than 50% of the time and does not interfere with lying, sitting, standing and/or walking

3 – Moderate: Dystonia more than 50% of the time and/or dystonia that interferes with lying, sitting, standing and/or walking

4 – Severe: Dystonia more than 50% of the time and/or dystonia that prevents sitting in a standard wheelchair, standing and/or walking (e.g. requires more than standard head rest for seating)

* Unable to assess neck movements

NECK: .....

TRUNK: Signs of trunk dystonia include pulling of the trunk into any plane of motion: extension, flexion, lateral flexion or rotation

0 – Absence of trunk dystonia

1 – Slight: Pulling less than 10% of the time and does not interfere with lying, sitting, standing and/or walking

2 – Mild: Dystonia less than 50% of the time and does not interfere with lying, sitting, standing and/or walking

3 – Moderate: Dystonia more than 50% of the time and/or dystonia that interferes with lying, sitting, standing and/or walking

4 – Severe: Dystonia more than 50% of the time and/or dystonia that prevents sitting in a standard wheelchair, standing and/or walking (e.g. requires adapted seating system to control posture, such as ASIS bar)

* Unable to assess trunk movements

TRUNK: .....

UPPER EXTREMITIES: Signs of dystonia in the upper extremities includes: sustained muscle contractions causing abnormal posturing of the upper extremities

0 – Absence of upper extremity dystonia

1 – Slight: Dystonia less than 10% of the time and does not interfere with normal positioning and/or functional activities

2 – Mild: Dystonia less than 50% of the time and does not interfere with normal positioning and/or functional activities

3 – Moderate: Dystonia more than 50% of the time and/or dystonia that interferes with normal positioning and/or upper extremity function

4 – Severe: Dystonia more than 50% of the time and/or dystonia that prevents normal positioning and/or upper extremity function (e.g. arms restrained in a wheelchair to prevent injury)

* Unable to assess upper extremity movements

LEFT UPPER EXTREMITY: .....

RIGHT UPPER EXTREMITY: .....

LOWER EXTREMITIES: Signs of dystonia in the lower extremities include: sustained muscle contractions causing abnormal posturing of the lower extremities

0 – Absence of lower extremity dystonia

1 – Slight: Dystonia less than 10% of the time and does not interfere with normal positioning and/or functional activities

2 – Mild: Dystonia less than 50% of the time and does not interfere with normal positioning and/or functional activities

3 – Moderate: Dystonia more than 50% of the time and/or dystonia that interferes with normal positioning and/or lower extremity weight bearing and/or function

4 – Severe: Dystonia more than 50% of the time and/or dystonia that prevents normal positioning and/or lower extremity weight bearing and/or function (e.g. cannot maintain standing owing to severe dystonia at ankles)

* Unable to assess lower extremity movements

LEFT LOWER EXTREMITY: .....

RIGHT LOWER EXTREMITY: .....

Total score: ..... **Rater's Initials:** .....

UNIFIED DYSTONIA RATING SCALE (UDRS)

Comella CL, Leurgans S, Wuu J, Stebbins GT, Chmura T & Dystonia Study Group (2002)

1. DURATION FACTOR

0 None

0.5 Occasional (<25% of the time) predominantly submaximal

1.0 Occasional (<25% of the time) predominantly maximal

1.5 Intermittent (25-50% of the time) predominantly submaximal

2.0 Intermittent (25-50% of the time) predominantly maximal

2.5 Frequent (50-75% of the time) predominantly submaximal

3.0 Frequent (50-75% of the time) predominantly maximal

3.5 Constant (>75% of the time) predominantly submaximal

4.0 Constant (>75% of the time) predominantly maximal

2. MOTOR SEVERITY FACTOR

EYES AND UPPER FACE

0 None

1 Mild: Increased blinking and/or slight forehead wrinkling (≤25% maximal intensity)

2 Moderate: eye closure without squeezing and/or pronounced forehead wrinkling (>25% but ≤50% maximal intensity)

3 Severe: eye closure with squeezing, able to open eyes within 10 seconds and/or marked forehead wrinkling (>50% but ≤75% maximal)

4 Extreme: Eye closure with squeezing, unable to open eyes within 10 seconds and/or intense forehead wrinkling (≥75% maximal intensity)

LOWER FACE

0 None

1 Mild: grimacing of lower face with minimal distortion of mouth (≤25% maximal)

2 Moderate: grimacing of lower face with moderate distortion of mouth (>25% but ≤50% maximal)

3 Severe: marked grimacing with severe distortion of mouth (>50% but ≤75% maximal)

4 Extreme: intense grimacing with extreme distortion of mouth (>75% maximal)

JAW AND TONGUE

0 None

1 Mild: jaw opening and/or tongue protrusion ≤25% of possible range or forced jaw clenching without bruxism

2 Moderate: jaw opening and/or tongue protrusion >25% but ≤50% of possible range or forced jaw clenching with mild bruxism secondary to dystonia

3 Severe: jaw opening and/or tongue protrusion >50% but ≤75% of possible range or forced jaw clenching with pronounced bruxism secondary to dystonia

4 Extreme: jaw opening and/or tongue protrusion >75% of possible range or forced jaw clenching with inability to open mouth

LARYNX

0 None

1 Mild: barely detectable hoarseness and/or choked voice and/or occasional voice breaks

2 Moderate: obvious hoarseness and/or choked voice and/or occasional voice breaks

3 Severe: marked hoarseness and/or choked voice and/or occasional voice breaks

4 Extreme: unable to vocalise

NECK

0 None

1 Mild: movement of head from neutral position ≤25% of possible normal range

2 Moderate: movement of head from neutral position >25% but ≤50% of possible normal range

3 Severe: movement of head from neutral position >50% but ≤75% of possible normal range

4 Extreme: movement of head from neutral position >75% of possible normal range

SHOULDER & PROXIMAL ARM (RIGHT & LEFT)

0 None

1 Mild: movement of shoulder or upper arm ≤25% of possible normal range

2 Moderate: movement of shoulder or upper arm >25% but ≤50% of possible normal range

3 Severe: movement of shoulder or upper arm >50% but ≤75% of possible normal range

4 Extreme: movement of shoulder or upper arm >75% of possible normal range

DISTAL ARM & HAND INCLUDING ELBOW (RIGHT & LEFT)

0 None

1 Mild: movement of distal arm or hand ≤25% of possible normal range

2 Moderate: movement of distal arm or hand >25% but ≤50% of possible normal range

3 Severe: movement of distal arm or hand >50% but ≤75% of possible normal range

4 Extreme: movement of distal arm or hand >75% of possible normal range

PELVIS AND PROXIMAL LEG (RIGHT & LEFT)

0 None

1 Mild: tilting of pelvis or movement of proximal leg or hip ≤25% of possible normal range

2 Moderate: tilting of pelvis or movement of proximal leg or hip >25% but ≤50% of possible normal range

3 Severe: tilting of pelvis or movement of proximal leg or hip >50% but ≤75% of possible normal range

4 Extreme: tilting of pelvis or movement of proximal leg or hip >75% of possible normal range

DISTAL LEG AND FOOT INCLUDING KNEE (RIGHT & LEFT)

0 None

1 Mild: movements of distal leg or foot ≤25% of possible normal range

2 Moderate: movements of distal leg or foot >25% but ≤50% of possible normal range

3 Severe: movements of distal leg or foot >50% but ≤75% of possible normal range

4 Extreme: movements of distal leg or foot >75% of possible normal range

TRUNK

0 None

1 Mild: bending of trunk ≤25% of possible normal range

2 Moderate: bending of trunk >25% but ≤50% of possible range

3 Severe: bending of trunk >50% but ≤75% of possible range

4 Extreme: bending of trunk >75% of possible range

Duration score:...../56 **Severity score:**...../56 **Total score:**...../112

MOVEMENT DISORDER – CHILDHOOD RATING SCALE (MD-CRS)

Battini R, Sgandurra G, Petacchi E, Guzzetta A, DiPietro R, Giannini MT, Leuzzi V, Mercuri E & Cioni G. (2008)

PART 1: GENERAL ASSESSMENT

A: MOTOR FUNCTION

1. HEAD CONTROL (video > 1 minute)
0 – normal
1 – mildly affected by MD and/or control > 1 min
2 – mod affected by MD and/or control between 30 sec and 1 min
3 – severely affected by MD and/or control < 30 secs
4 – absent

2. SITTING POSITION (video > 1 minute)
0 – normal
1 – mildly affected by MD and/or control > 1min
2 – mod affected by MD and/or control between 30 sec and 1 min
3 – severely affected by MD and/or control < 30 secs
4 – absent

3. STANDING POSITION (video > 1 minute)
0 – normal
1 – maintained for > 1 min, even with abnormal posture
2 – maintained for > 30 sec and < 1 minute, even with abnormal posture
3 – maintained for < 30 seconds, or long periods with support
4 – absent or for brief periods with support

4. WALKING (video > 1 minute, walk 5m)
0 – normal
1 – walks alone > 5m, even with abnormal posture
2 – walks alone < 5m, even with abnormal posture or needs support
3 – a few steps with support, with very abnormal posture
4 – absent

5. REACHING (video grasp/release of cubes)
0 – normal
1 – possible for age appropriate tasks, even if abnormal
2 – mod abnormal, only possible to approach object
3 – severely abnormal, only inefficient efforts
4 – absent

6. GRASPING (video grasp/release of cubes)
0 – normal
1 – mildly abnormal, pincer grasp possible
2 – mod abnormal, only possible for cubes close to hand
3 – severely abnormal, only inefficient efforts
4 – absent

7. HANDWRITING (video handwriting)
0 – normal
1 – mild difficulties, but readable
2 – mod difficulty, not completely readable
3 – severe difficulties, not readable
4 – absent, functional grasp of pen not possible

B. ORAL/VERBAL FUNCTION

1. SWALLOWING
0 – normal
1 – occasional dysphagia
2 – frequent dysphagia, swallowing difficulties
3 – swallowing solid foods not possible
4 – severe difficulty swallowing semi solids and liquids

2. DROOLING
0 – absent
1 – occasional
2 – mild
3 – moderate
4 – severe

3. LANGUAGE (video speech – e.g. name)
0 – normal
1 – mild dysarthria, speech comprehensible
2 – mod dysarthria, speech not fully comprehensible
3 – severe dysarthria, speech not comprehensible
4 – complete/almost complete anarthria

C. SELF-CARE

1. DRESSING (video dressing)
0 – complete autonomy
1 – min assistance (not fully efficient but independent)
2 – assistance in some tasks (shoes, buttons)
3 – partially dependent (co-operates in self-dressing)
4 – totally dependent

2. SELF-FEEDING
0 – complete autonomy (uses cutlery)
1 – min assistance (not fully efficient but independent)
2 – assistance in some tasks (using knife)
3 – partially dependent (uses hands to eat)
4 – totally dependent

3. PERSONAL CARE
0 – complete autonomy
1 – min assistance (not fully efficient but independent)
2 – assistance in some tasks (bathing)
3 – partially dependent (co-operates in teeth)
4 – totally dependent

D. ATTENTION/ALERTNESS

1. ATTENTION/ALERTNESS DURING THE OBSERVATION
0 – constantly alert, answers all age and mental adequate questions
1 – sometimes (1/3 of observation) inattentive
2 – often (2/3 of observation) inattentive
3 – attention needs to be constantly drawn by examiner
4 – constantly a scarce reaction to external stimuli

2. ATTENTION/ALERTNESS AT HOME
0 – constantly alert during day, good response
1 – sometimes (1/3 of day) inattentive
2 – often (2/3 of day) hypo-attentive
3 – child constantly sleepy, even during day
4 – constantly a scarce reaction to external stimuli

PART 2: MOVEMENT DISORDER ASSESSMENT

MD SEVERITY

1. EYE AND PERIORBITAL REGION (video at rest and during movement; fixate and follow visual stimulus)

0 – MD absent

1 – MD present only at rest

2 – MD present during 1 or some tasks for region examined and/or involves 1 or 2 other regions

3 – MD present during 1 or some tasks for region examined and involves >3 other regions

4 – MD present during all tasks for region examined and/or involves 3 or more other regions, making completion impossible

2. FACE (video at rest and during movement, smile, open and close eyes)

0 – MD absent

1 – MD present only at rest

2 – MD present during 1 or some tasks for region examined and/or involves 1 or 2 other regions

3 – MD present during 1 or some tasks for region examined and involves >3 other regions

4 – MD present during all tasks for region examined and/or involves 3 or more other regions, making completion impossible

3. TONGUE AND PERIORAL REGION (video at rest and during movement, stick out tongue and move in all directions)

0 – MD absent

1 – MD present only at rest

2 – MD present during 1 or some tasks for region examined and/or involves 1 or 2 other regions

3 – MD present during 1 or some tasks for region examined and involves >3 other regions

4 – MD present during all tasks for region examined and/or involves 3 or more other regions, making completion impossible

4. NECK (video at rest and during movement, dressing and sitting)

0 – MD absent

1 – MD present only at rest

2 – MD present during 1 or some tasks for region examined and/or involves 1 or 2 other regions

3 – MD present during 1 or some tasks for region examined and involves >3 other regions

4 – MD present during all tasks for region examined and/or involves 3 or more other regions, making completion impossible

5. TRUNK (video at rest and during movement, dressing and sitting)

0 – MD absent

1 – MD present only at rest

2 – MD present during 1 or some tasks for region examined and/or involves 1 or 2 other regions

3 – MD present during 1 or some tasks for region examined and involves >3 other regions

4 – MD present during all tasks for region examined and/or involves 3 or more other regions, making completion impossible

6. UPPER LIMBS (video at rest and during movement, dressing, sitting, moving cubes and writing)

0 – MD absent

1 – MD present only at rest

2 – MD present during 1 or some tasks for region examined and/or involves 1 or 2 other regions

3 – MD present during 1 or some tasks for region examined and involves >3 other regions

4 – MD present during all tasks for region examined and/or involves 3 or more other regions, making completion impossible

7. LOWER LIMBS (video at rest and during movement, dressing and sitting and standing/walking)

0 – MD absent

1 – MD present only at rest

2 – MD present during 1 or some tasks for region examined and/or involves 1 or 2 other regions

3 – MD present during 1 or some tasks for region examined and involves >3 other regions

4 – MD present during all tasks for region examined and/or involves 3 or more other regions, making completion impossible

CLASSIFICATION OF MD	PREVALENT MD	OTHER MD
Hypokinetic-rigid	☐	☐
Chorea/ballism	☐	☐
Dystonia/athetosis	☐	☐
Myoclonus	☐	☐
Tic	☐	☐
Tremor	☐	☐

MOVEMENT DISORDER – CHILDHOOD RATING SCALE 0-3 (MD-CRS 0-3)

Battini R, Guzzetta A, Sgandurra G, DiPietro R, Petacchi E, Mercuri E, Giannini MT, Leuzzi V, & Cioni G. (2009)

PART 1: GENERAL ASSESSMENT

A: MOTOR FUNCTION

1. HEAD CONTROL
0 – Normal
1 – Mildly affected by MD and/or control > 30 sec
2 – Mod affected by MD and/or control between 15 sec and 30 sec
3 – Severely affected by MD and/or control < 15 secs
4 – Absent

2. SITTING POSITION
0 – Normal
1 – Mildly affected by MD and/or control > 30 sec
2 – Mod affected by MD and/or control between 15 sec and 30 sec
3 – Severely affected by MD and/or control < 15 secs
4 – Absent

3. STANDING POSITION
0 – Normal
1 – Maintained for > 30 sec, even with abnormal posture
2 – Maintained for > 15 sec and < 30 sec, even with abnormal posture
3 – Maintained for < 15 sec, or long periods with support
4 – Absent or for brief periods with support

4. WALKING
0 – Normal
1 – Walks alone > 3m, even with abnormal posture
2 – Walks alone < 3m, even with abnormal posture or needs support
3 – A few steps with support, with very abnormal posture
4 – Absent

5. REACHING
0 – Normal
1 – Possible for age appropriate tasks, even if abnormal
2 – Mod abnormal, only possible to approach object
3 – Severely abnormal, only inefficient efforts
4 – Absent

6. GRASPING
0 – Normal
1 – Mildly abnormal, pincer grasp possible
2 – Mod abnormal, only possible for rattle close to hand
3 – Severely abnormal, only inefficient efforts
4 – Absent

B. ORAL/VERBAL FUNCTION

1. SWALLOWING
0 – Normal
1 – Occasional dysphagia
2 – Frequent dysphagia, swallowing difficulties
3 – Swallowing solid foods not possible
4 – Severe difficulty swallowing semi solids and liquids

2. DROOLING
0 – Absent
1 – Occasional
2 – Mild
3 – Moderate
4 – Severe

C. ATTENTION/ALERTNESS

1. ATTENTION/ALERTNESS DURING THE OBSERVATION
0 – Constantly alert, answers all age and mental adequate questions
1 – Sometimes (1/3 of observation) inattentive
2 – Often (2/3 of observation) inattentive
3 – Attention needs to be constantly drawn by examiner
4 – Constantly a scarce reaction to external stimuli

2. ATTENTION/ALERTNESS AT HOME
0 – Constantly alert during day, good response
1 – Sometimes (1/3 of day) inattentive
2 – Often (2/3 of day) hypo-attentive
3 – Child constantly sleepy, even during day
4 – Constantly a scarce reaction to external stimuli

PART 2: MOVEMENT DISORDER ASSESSMENT

MD SEVERITY

1. EYE AND PERIORBITAL REGION
0 – MD absent
1 – MD is intermittent
2 – MD is constant

2. FACE
0 – MD absent
1 – MD is intermittent
2 – MD is constant

3. TONGUE AND PERIORAL REGION
0 – MD absent
1 – MD is intermittent
2 – MD is constant

4. NECK
0 – MD absent
1 – MD is intermittent
2 – MD is constant

5. TRUNK
0 – MD absent
1 – MD is intermittent
2 – MD is constant

6. UPPER LIMBS
0 – MD absent
1 – MD is intermittent
2 – MD is constant

7. LOWER LIMBS
0 – MD absent
1 – MD is intermittent
2 – MD is constant

CLASSIFICATION OF MD	PREVALENT MD	OTHER MD
Hypokinetic-rigid	☐	☐
Chorea/ballism	☐	☐
Dystonia/athetosis	☐	☐
Myoclonus	☐	☐
Tic	☐	☐
Tremor	☐	☐

THE DYSKINESIA IMPAIRMENT SCALE (DIS)

Monbaliu E, Ortibus E, de Cat J, Dan B, Heyman L, Prinzie P, de Cock P & Feys H. (2012)

DYSKINETIC CEREBRAL PALSY: is characterised by involuntary, uncontrolled recurring, occasionally stereotyped movements, in which the primitive reflex patterns predominate and the muscle tone is varying. It is further subdivided into dystonia and choreoathetosis

DYSTONIA: in cerebral palsy is predominated by abnormal postures (may give impression of hypokinesia) and muscle tone that is fluctuating (but with easily elicitable tone increase). Characteristics are involuntary movements, distorted voluntary movements and abnormal postures due to sustained muscle contractions.

CHOREOATHETOSIS: in cerebral palsy is predominated by hyperkinesia and tone fluctuating (but mainly decreased). Chorea means rapid involuntary jerky, often fragmented movements. Athetosis means slower, constantly changing writhing or contorting movements.

REGION	DYSTONIA	CHOREOATHETOSIS
Eye	Dystonia around **eyes, eyelids, eyebrow, forehead**: e.g. sustained muscle contractions (blepharospasms) around eyes and/or eyelids (open/closed) and/or forced eye movement deviations, e.g. during eye tracking movement or fixation	Choreoathetosis around eyes, **eyelids, eyebrows, forehead**: e.g. constantly, fragmented movements around eyes and/or blinking eyelid (open/closed) and/or variable (saccadic) eye movements, e.g. during eye tracking movement or fixation
Mouth	Dystonia around **lips, jaw, cheeks, tongue**: e.g. sustained muscle contraction resulting in grimacing movement, clenched or deviated jaw, forced open mouth and/or forceful tongue thrust	Choreoathetosis around **lips, jaw, cheeks, tongue**: e.g. constantly changing, fragmented movements in the lower face such as grimacing, mouth movements and tongue protrusion movements
Neck	Dystonia in **neck**: sustained muscle contraction resulting in pulling neck movements and/or posture into any plane of motion – ext, flex, lat. flex and rot	Choreoathetosis in **neck**: e.g. constantly changing or fragmented neck movements (wagging) into any plane of motion - ext, flex, lat. flex and rot
Trunk	Dystonia in **trunk**: e.g. sustained muscle contraction resulting in pulling trunk movements and/or posture into any plane of motion – ext, flex, lat. flex and rot	Choreoathetosis in **trunk**: e.g. constantly changing fragmented or contorting trunk movements (wagging) into any plane of motion - ext, flex, lat. flex and rot
Arm (proximal)	Dystonia in the **shoulder girdle, upper arm, elbow**: e.g. sustained muscle contractions causing abnormal posturing, involuntary and/or distorted voluntary movements of the proximal arm	Choreoathetosis in the **shoulder girdle, upper arm, elbow**: e.g. constantly changing fragmented or contorting movements of proximal arm: jerky, stormy (choreo) and/or wriggling, contorting (athetosis)
Arm (distal)	Dystonia in **forearm, wrist, hand**: e.g. sustained muscle contractions causing abnormal posturing, involuntary and/or distorted voluntary movements of distal arm	Choreoathetosis in **forearm, wrist, hand**: e.g. constantly changing fragmented or contorting movements of distal arm: jerky, stormy (choreo) and/or wriggling, contorting (athetosis)
Leg (proximal)	Dystonia in **hip girdle, upper leg, knee**: e.g. sustained muscle contractions causing abnormal posturing and/or distorted voluntary movements of proximal leg	Choreoathetosis in **hip girdle, upper leg, knee**: e.g. constantly changing fragmented or contorted movements of proximal leg: jerky, stormy (chorea) and/or wriggling, contorting (athetosis)
Leg (distal)	Dystonia in **lower leg, ankle, foot**: e.g. sustained muscle contractions causing abnormal posturing and/or distorted voluntary movements of distal leg	Choreoathetosis in **lower leg, ankle, foot**: e.g. constantly changing fragmented or contorted movements of distal leg: jerky, stormy (chorea) and/or wriggling, contorting (athetosis)

DURATION FACTOR	AMPLITUDE FACTOR
0 = D/CA is absent	0 = D/CA is absent
1 = D/CA is occasionally present (< 10%)	1 = in small ROM (< 10%)
2 = D/CA is frequently present (≥ 10 – < 50%)	2 = D/CA in moderate ROM (≥ 10 – < 50%)
3 = D/CA is mostly present (≥ 50 – < 90%)	3 = D/CA in submaximal ROM (≥ 50 – < 90%)
4 = D/CA is always present (≥ 90%)	4 = D/CA in maximal ROM (≥ 90%)

DYSTONIA REPORTING FOR DYSKINESIA IMPAIRMENT SCALE (DIS)

REGION	ACTION			REST		
	SINGLE ACTIVITY	DURATION FACTOR	AMPLITUDE FACTOR	SINGLE POSITION	DURATION FACTOR	AMPLITUDE FACTOR
Eye	Eye tracking	0-1-2-3-4	0-1-2-3-4	Sitting	0-1-2-3-4	0-1-2-3-4
	Eye blinking	0-1-2-3-4	0-1-2-3-4			
Mouth	Mouth open/closed	0-1-2-3-4	0-1-2-3-4	Sitting	0-1-2-3-4	0-1-2-3-4
	Speech	0-1-2-3-4	0-1-2-3-4			
Neck	Lateroflexion right/left	0-1-2-3-4	0-1-2-3-4	Sitting	0-1-2-3-4	0-1-2-3-4
	Rotation right/left	0-1-2-3-4	0-1-2-3-4			
Trunk	Active sitting position	0-1-2-3-4	0-1-2-3-4	Sitting	0-1-2-3-4	0-1-2-3-4
	Forward flexion	0-1-2-3-4	0-1-2-3-4			
Right arm proximal	Arm abduction	0-1-2-3-4	0-1-2-3-4	Sitting	0-1-2-3-4	0-1-2-3-4
	Grasp and move a pen	0-1-2-3-4	0-1-2-3-4			
Left arm proximal	Arm abduction	0-1-2-3-4	0-1-2-3-4	Sitting	0-1-2-3-4	0-1-2-3-4
	Grasp and move a pen	0-1-2-3-4	0-1-2-3-4			
Right arm distal	Grasp and move a cup	0-1-2-3-4	0-1-2-3-4	Sitting	0-1-2-3-4	0-1-2-3-4
	Grasp and move a pen	0-1-2-3-4	0-1-2-3-4			
Left arm distal	Grasp and move a cup	0-1-2-3-4	0-1-2-3-4	Sitting	0-1-2-3-4	0-1-2-3-4
	Grasp and move a pen	0-1-2-3-4	0-1-2-3-4			
Right leg proximal	Rolling	0-1-2-3-4	0-1-2-3-4	Lying	0-1-2-3-4	0-1-2-3-4
	Standing	0-1-2-3-4	0-1-2-3-4			
Left leg proximal	Rolling	0-1-2-3-4	0-1-2-3-4	Lying	0-1-2-3-4	0-1-2-3-4
	Standing	0-1-2-3-4	0-1-2-3-4			
Right leg distal	Rolling	0-1-2-3-4	0-1-2-3-4	Lying	0-1-2-3-4	0-1-2-3-4
	Heel/toe raising	0-1-2-3-4	0-1-2-3-4			
Left leg distal	Rolling	0-1-2-3-4	0-1-2-3-4	Lying	0-1-2-3-4	0-1-2-3-4
	Heel/toe raising	0-1-2-3-4	0-1-2-3-4			

Total score: .....

CHOREOATHETOSIS REPORTING FOR DYSKINESIA IMPAIRMENT SCALE (DIS)

REGION	ACTION			REST		
	SINGLE ACTIVITY	DURATION FACTOR	AMPLITUDE FACTOR	SINGLE POSITION	DURATION FACTOR	AMPLITUDE FACTOR
Eye	Eye tracking	0-1-2-3-4	0-1-2-3-4	Sitting	0-1-2-3-4	0-1-2-3-4
	Eye blinking	0-1-2-3-4	0-1-2-3-4			
Mouth	Mouth open/closed	0-1-2-3-4	0-1-2-3-4	Sitting	0-1-2-3-4	0-1-2-3-4
	Speech	0-1-2-3-4	0-1-2-3-4			
Neck	Lateroflexion right/left	0-1-2-3-4	0-1-2-3-4	Sitting	0-1-2-3-4	0-1-2-3-4
	Rotation right/left	0-1-2-3-4	0-1-2-3-4			
Trunk	Active sitting position	0-1-2-3-4	0-1-2-3-4	Sitting	0-1-2-3-4	0-1-2-3-4
	Forward flexion	0-1-2-3-4	0-1-2-3-4			
Right arm proximal	Arm abduction	0-1-2-3-4	0-1-2-3-4	Sitting	0-1-2-3-4	0-1-2-3-4
	Grasp and move a pen	0-1-2-3-4	0-1-2-3-4			
Left arm proximal	Arm abduction	0-1-2-3-4	0-1-2-3-4	Sitting	0-1-2-3-4	0-1-2-3-4
	Grasp and move a pen	0-1-2-3-4	0-1-2-3-4			
Right arm distal	Grasp and move a cup	0-1-2-3-4	0-1-2-3-4	Sitting	0-1-2-3-4	0-1-2-3-4
	Grasp and move a pen	0-1-2-3-4	0-1-2-3-4			
Left arm distal	Grasp and move a cup	0-1-2-3-4	0-1-2-3-4	Sitting	0-1-2-3-4	0-1-2-3-4
	Grasp and move a pen	0-1-2-3-4	0-1-2-3-4			
Right leg proximal	Rolling	0-1-2-3-4	0-1-2-3-4	Lying	0-1-2-3-4	0-1-2-3-4
	Standing	0-1-2-3-4	0-1-2-3-4			
Left leg proximal	Rolling	0-1-2-3-4	0-1-2-3-4	Lying	0-1-2-3-4	0-1-2-3-4
	Standing	0-1-2-3-4	0-1-2-3-4			
Right leg distal	Rolling	0-1-2-3-4	0-1-2-3-4	Lying	0-1-2-3-4	0-1-2-3-4
	Heel/toe raising	0-1-2-3-4	0-1-2-3-4			
Left leg distal	Rolling	0-1-2-3-4	0-1-2-3-4	Lying	0-1-2-3-4	0-1-2-3-4
	Heel/toe raising	0-1-2-3-4	0-1-2-3-4			

Total score: .....

11. APPENDIX 2

SURVEY RESULTS: DYSKINESIA IN CEREBRAL PALSY: ITS IDENTIFICATION, CLASSIFICATION AND MEASUREMENT

A survey was developed that aimed to investigate clinicians' knowledge, and perceived barriers, relating to identification and measurement of dyskinesia (dystonia and choreoathetosis) in children with cerebral palsy and explore educational needs regarding improving identification and assessment of dyskinesia.

The online, anonymous, self-report survey was distributed via clinical and professional networks across Australia. Hospital and community clinicians including medical doctors and allied health clinicians involved in the treatment of children with cerebral palsy were targeted.

The raw results of the survey are contained in this section. These results have served to inform the need for and the content of the toolkit and the knowledge transfer strategies that will be implemented with clinicians. In total 170 Australian clinicians attempted the survey with n=163 completed surveys to analyse. Not all respondents completed every question.

SECTION 1

Information regarding profession and current work place/s, educational qualifications, and experience working with children with cerebral palsy.

Table 1: Demographic data

FACTOR	CATEGORIES	*n* RESPONDENTS *(n=163)*	%
Profession	Physiotherapist	95	58.2
	Occupational Therapist	39	23.9
	Rehab Specialist	11	6.7
	Rehab Specialist / Paediatrician	2	1.2
	Neurologist	1	0.6
	Paediatric Neurologist	3	1.8
	Neurologist / Paediatrician	1	0.6
	Orthopaedic Surgeon	3	1.8
	Speech Pathologist	4	2.4
	Nurse	2	1.2
	Social Worker	1	0.6
	Neuropsychologist	1	0.6
	Researcher (Physio or OT)	3	
Education level	Diploma	4	2.4
	Bachelor	85	52.1
	Masters	60	36.8
	Doctorate	13	8
	Other	1	0.6
Years of experience with children with cerebral palsy	< 1 year	5	3.1
	1 to 4 years	22	13.5
	5 to 10 years	48	29.4
	> 10 years	88	54
Proportion of caseload children with cerebral palsy	< 10%	20	12.3
	10 – 24%	25	15.3
	25 – 49%	28	17.2
	50 – 74%	42	25.8
	> 75%	48	29.4
Current workplace	Tertiary hospital	63	38.7
	Not for profit organisation	57	35
	Private practice	17	10.4
	Hospital	15	9.2
	Community health	14	8.6
	Government organisation	11	6.7
	University	4	2.5
	Schools (mainstream x5 / supported x6)	8	4.9
	Research facility	7	4.3

SECTION 2

Clinicians' knowledge about dyskinesia in cerebral palsy. Investigated clinicians' ability to identify and differentiate between different movement disorders and what they saw as the barriers to accurate identification.

Table 2: Knowledge about dyskinesia

FACTOR	CATEGORIES	*n* RESPONDENTS	% (n)
Differentiate / describe MD	Yes	150	95 (157)
	No	7	
	N/A	0	
How identify dyskinesia	Clinical judgement/observation	136	91 (149)
	MDT Assessment	83	56
	Video analysis	59	39
	Assessment tool/class system[a]	88	59
	Other	2	
Movement disorders identified	Spasticity	148	99 (149)
	Dystonia	139	93
	Chorea	51	34
	Athetosis	69	46
	Ballismus	12	8
	Ataxia	86	58
Barriers to accurate identification	Limited access to training	64	40.8 (157)
	Limited training availability	64	40.8
	Lack of time	40	25.5
	Limited applicability to caseload	23	14.6
	Limited knowledge – what look like	60	38.2
	N/A to current role	7	4.5
	Lack of clinically applicable tools	79	50.3
	Limited confidence to identify	70	44.6
	Other[b]	17	10.8

a. Which assessment / classification tool(s):

HAT n=56

BADS n=31

BFMDRS n=12

DIS n=10

GDS n=1

ASAS n=19

MAS n=5

MTS n=8

AROM n=1

SCPE n=1

International classification of movement disorders n=1

CP registry Ax n=1

GMFCS/MACS/CFCS n=2

Type & topography n=1

CP description form n=2

Observational gait analysis n=1

b. Barriers to accurate identification of dyskinesia in children with CP?

- Presence of mixed movement disorders, difficult to distinguish between.
- Use of the ASAS clinical description form relies on you knowing what to look for.
- Would like to use DIS more but hard due to time.
- Assessments that are clinically appropriate such as DIS take a long time to complete, also need to be disciplined about making sure video is taken routinely.
- Suitable charts to guide initial examinations and functional assessments for identification.
- Poor consensus between professionals makes it confusing.
- I don't see a barrier for myself.
- For me, it is not part of my role or expertise.
- Large proportion of clients present with combined types of dyskinesia, and spasticity, can make it difficult to assess/clearly determine an accurate identification.
- Lack of national and international definitions, neurologists and paeds and rehab persons often label the same movement problem differently.
- Working with infants: dystonia becomes evident with goal directed movement and fluctuates.
- There are standardised tools to identify and measure dyskinesia but they are often complicated and time-consuming to use in the regular clinical setting.
- Ambiguity and inconsistency in the accepted clinical phenotypes.
- I often have clinical discussions with other therapists (i.e. MDT) about what is dystonia. I find there is a poor understanding of dystonia and what this manifests into in a clinical presentation.
- Lack of understanding of the different phenotypes and confusion with definitions.
- Tendency to just look for spasticity.
- Occasional discrepancies or differences between various clinicians' definitions of and distinctions between dystonia, athetosis and chorea still exist.
- N/A.
- The difficulty clinically when part of other tone issues that may mask or make identification clear.
- Time management to complete formal dyskinesia Ax.
- Variability in presentation and report from family or other people involved.
- Variations in professional opinion and use of terms.

SECTION 3

Focused specifically on clinical knowledge of the various tools available to measure or quantify dyskinesia in children with cerebral palsy. Survey respondents were asked to indicate which tools they had knowledge of, how familiar they were with the tools, how useful they found them, what area of clinical practice they used them for and what type of training is required for their accurate use.

Table 3: Tools available to measure/quantify dyskinesia

KEY: N (%)		BADS	BFMDRS	DIS	UDRS	MD-CRS / MD-CRS 0-3	NIL
Knowledge of tool (10%)	Other[c] = 16	103 (63.2)	42 (25.8)	53 (32.5)	16 (9.8)	13 (8) 8 (5)	38 (23.3)
Familiarity	Very familiar, use frequently (18.9%)	25 (24.3)	11 (26.2)	6 (11.3)	0	1 (7.7)	Mean: 13.9%
	Somewhat familiar, used few times (48.5%)	57 (55.3)	14 (33.3)	28 (52.8)	8 (50)	3 (23.1)	Mean: 42.9%
	Not familiar, heard of but don't use it (32.6%)	21 (20.4)	17 (40.5)	18 (34)	8 (50)	10 (77)	Mean: 44.4%
Training	Training required for accurate use (89.9%)	96 (93.2)	36 (85.7)	48 (90.6)	12 (75)	12 (92.3)	Mean: 87.4%
Type training	Read journal article	26 (25.2)	12 (28.6)	15 (28.3)	4 (25)	2 (15.4)	Mean: 23.9%
	Clinical training by experienced clinicians	77 (74.8)	31 (73.8)	33 (62.3)	7 (43.75)	6 (46.2)	Mean: 60.2%
	Manual	48 (46.6)	15 (35.7)	23 (43.4)	5 (31.25)	3 (23.1)	Mean: 39.1%
	Course	31 (30.1)	17 (40.5)	26 (49.1)	5 (31.25)	5 (38.5)	Mean: 37.9%
	Other (videos)	17 (16.5)	5 (11.9)	6 (11.3)	3 (18.75)	0	
Usefulness (196)	Extremely useful 12.8%	10 (9.7)	3 (7.1)	11 (20.7)	0	1 (7.7)	Mean: 9.05%
	Somewhat useful 69.4%	74 (71.8)	20 (47.6)	30 (56.6)	7 (43.75)	5 (38.5)	Mean: 51.6%
	Not useful at all 17.8%	8 (7.8)	11 (26.2)	6 (11.3)	5 (31.25)	5 (38.5)	Mean: 23.0%
Purpose	Routine therapy and Ax (15% of total 227 scales)	22 (21.4)	4 (9.5)	7 (13.2)	0	1 (7.7)	Mean: 10.4%
	Routine MDT Ax 24.7%	32 (31.1)	13 (31)	10 (18.9)	0	1 (7.7)	Mean: 17.7%
	Med trials 24.7%	31 (30.1)	9 (21.4)	13 (24.5)	1 (6.25)	2 (15.4)	Mean: 19.5%
	DBS 29.1%	27 (26.2)	23 (54.8)	12 (28.3)	2 (12.5)	2 (15.4)	Mean: 27.4%
	ITB 27.75%	36 (35)	14 (33.3)	11 (20.7)	0	2 (15.4)	Mean: 20.9%
	Research 20.3%	21 (20.4)	11 (26.2)	9 (17)	3 (18.75)	2 (15.4)	Mean: 19.5%
	Don't use it 30%	22 (21.4)	14 (33.3)	16 (30.2)	9 (56.3)	7 (54)	Mean: 39%

c. Other tools:

HAT = 16

ASAS = 3

GDS = 2

WA CP description form = 1

SECTION 4

The final section of the survey investigated clinicians educational and clinical needs regarding dyskinesia identification and measurement. Respondents were asked about any formal and informal training they had attended in this area and whether a greater understanding of dyskinesia and movement disorders and their measurement would be clinically useful.

Table 4: Barriers to use of dyskinesia tools

FACTOR	CATEGORIES	RESPONDENTS *n* = 151	%
Barriers	Limited training opportunities	83	55
	Lack of time to learn	42	27.8
	Lack of confidence using available tools correctly/accurately	73	48.3
	Lack of time to complete assessments	60	39.7
	Limited applicability to total caseload	36	23.8
	Limited knowledge of the different tools that are available	60	39.7
	Limited knowledge which tools best suit different clinical presentations	68	45
	Tools not clinically meaningful or feasible	24	15.9
	My therapy team utilise tools for me as part of our full clinical assessment	15	9.9
	Other[d]	11	7.3

d. Other barriers:

- Not clinically meaningful when in community setting.
- Lack of refresher training available.
- I think DIS provides the most accurate information, but it has limited clinical utility due to the time taken to complete the assessment.
- Lack of clinical supervision.
- Current tools are not sensitive enough to show a clinically significant change when medication is being used to impact on dyskinesia such as Artane. Apart from GAS or function related outcome measures I don't find the current dystonia measures are adding enough to help us advise back to the hospital if the Artane should be continued. We have discussed a newer DIS but our senior clinician said that the research had not been completed yet so asked us to keep using the HAT.
- Buying scoring sheets/paper resources in order to use the scales (cost).
- Not usually requested in private practice.
- The tools need to be short, easy to administer by experienced clinicians who can also see utility over clinical impression or judgement.
- Not my role or expertise.
- Challenges with clinically meaningful and sensitive tools to accurately measure dyskinesia. Limited applicability to determine the impact of the impairment on function!
- Lack of confidence/limited knowledge is because there may not be guidelines that exist on how to use the tool.
- I am more familiar with the HAT. I would probably use some of the other tools if I was more familiar with them. I would use the tool that was most useful to me as a clinical tool in the context of therapy. Most of the tools seem to be qualitative and quantitative and do not focus on functional outcomes. I tend to use functional outcome measures more often in clinical practice, other than for initial assessment.
- If there was a child I was seeing, there would be more appropriate team members to carry out the assessment.
- Time to consolidate learning and implement new tools.
- Variability in presentation of children with dyskinesia and whether or not a one point in time assessment is clinically meaningful (parent reports can be vastly different).

Table 5.: Educational and clinical needs re. dyskinesia identification and measurement

FACTOR	CATEGORIES	RESPONDENTS *n* = 145	%
Formal training in dyskinesia identification	Yes[e]	54	37.2
	No	91	62.8
Training on specific measure?	Yes[f]	44	30.3
	No	101	69.7
Would a greater understanding of dyskinesia be useful for your current clinical role?	Extremely useful	60	41.4
	Very useful	51	35.2
	Somewhat useful	27	18.6
	Not very useful	7	4.8
	Not useful at all	0	
Would a greater understanding of the various tools available to identify and measure dyskinesia in children with cerebral palsy be useful for your current clinical role?	Extremely useful	55	38
	Very useful	49	34
	Somewhat useful	29	20
	Not very useful	11	7.6
	Not useful at all	0	
If a training package or 'toolkit' on identification and measurement of dyskinesia in children with cerebral palsy were developed, which format(s) would be of most benefit in your workplace:	Written information	67	46.2
	Video examples of MDs and how to assess	125	86.2
	Video training (examples of tools)	116	80
	Interactive workshops	83	57.2
	Webinars	87	60
	App – use in clinic to help identify and measure	76	52.4
	Other[g]		

e. Formal training in dyskinesia:

If you answered 'Yes', please provide some details of the training:

SUMMARY

DIS Course (16), Bobath course (9), Conferences (16), HAT Course (4), CPA in-services (5), In-services (14), Advanced course (1), Medical training (4).

f. Please provide details of the specific assessments/measures you have had training in:

Summary of comments: DIS training (20), UDRS (2), BADS (4), BFMDRS (2), HAT (6), ASAS (3), Conference workshops including AusACPDM, AACPDM, EACD (13), CPA training (2), In-services (2).

g. In a training package or 'toolkit' on identification and measurement of dyskinesia in children with cerebral palsy, which of the following would be of most benefit in your workplace:

- Great for orientation of new team members, benchmarking, updates.
- Networking with colleagues using the specific tools so that there is ease in communication in regards to questions about the tool that may arise.
- Manual with all information on all assessments in one place.
- My answer would be more for the purpose of being able to identify rather than assess common movement disorders.
- Relating assessment finding to clinical decision making.
- Targeted to infants.
- I am very interested in this whole endeavour, but some of my answers are personally pragmatic as I expect to retire in a couple of years.
- Sorry all above options are better than what we have now!

Please add any further information you feel may be helpful to improve the identification and assessment of dyskinesia in children with cerebral palsy:

- Written information is always a good adjunct but I find that videos are totally indispensable when learning about dyskinesia because it is a movement disorder that is 'seen' not felt or read about! We need to have ready access to example videos that exemplify the characteristics of each disorder.
- I think training in the epidemiology of dyskinetic cerebral palsy is interesting. So many clients come with a diagnosis of 'spastic quadriplegia' and with simple assessment it can be identified dyskinetic cerebral palsy is more appropriate.
- I miss being part of a team of skilled therapists with access to high quality PD and clinical supervision such as at Yooralla and Scope. Though working in a Key Worker model in Early Childhood intervention has given me skills in other areas, it has watered down professional skills and confidence.
- 'Hands on' training is necessary to observe the variability associated with dyskinesia and also to move clinicians on from the fixation with the Tardieu which may be falsely reassuring them about the issues of dyskinesia when the child has little or no (co-existing) spasticity. I feel strongly that this needs to be shown/demonstrated in an interactive workshop with patients with the various forms of dyskinetic movement disorders.
- One of the challenges is knowing how this identification will help clinical management. We need to improve consistency of classification before we can look at research on intervention approaches for different movement disorders. Currently it is difficult to justify too much time spent on assessment without any explicit benefit for the child in the short-term.
- Differentiation between chorea and athetosis is probably the most difficult to identify.
- Some of the recent paediatric neurology literature with involuntary movement disorders in children is extremely good, especially for definitions. Also, 'red flags' for when differential diagnoses should be considered and investigated while undergoing this process of identification and assessment of various movement disorders.
- I think apps are a valuable tool in current/future practice. I am currently doing a research project on evaluating an app for developing video-based exercise programs.
- Better assessment and discrimination of different types of dyskinesia (e.g. dystonia vs choreoathetosis) may help to be able to identify which patients will benefit from different treatments e.g. ITB, oral medications, DBS etc.

- Definitely more PD around cerebral palsy.
- It is more motivating to link identification of dyskinesia to specific treatment options or outcomes. Identification and labelling of dyskinesia without clinical reason just feels pedantic.
- I think in rural areas as staff are often isolated, confidence and knowledge in completing assessments is often low and there are not staff available who have completed the assessments regularly. I feel a web based or app based training that could be available during a session with a client would be really helpful. Thanks.
- Common terminology being used by *all* health practitioners would be extremely helpful. It will be great to have a widely available and utilised toolkit that puts everyone on the same page.
- For research purposes this might be useful but in practice its use is limited, still useful in certain specific areas but not very useful in practice. This is very medical model thinking!
- Need to see the movement disorder to then label it. Need consensus of neurologists, paediatrician's rehabilitation consultants and therapists in disability to accurately label the movement disorder. Need a new tool which is sensitive to change when trialling different treatments for dyskinesia.
- Neurologists seem to have a different understanding of some of our definitions and how to assess and manage...it would be in the best interest of the children that everyone involved in their care would use the same definition and they may also be interviewed for this purpose?
- Within my role I am not involved with the various tools available to identify and measure dyskinesia, but it would be useful to identify this as this form of movement disorder is impacting the child's ADL, carer burden etc. If I understand this better, I will also have a better understanding of the challenges patients and their carers' experience.
- The workshop by Dr Monbaliu I attended several years ago was very useful however the assessment tool he described was very lengthy and time consuming to apply, hence I have never used it.
- Clinical time and clinical feasibility for those children where it is important to differentiate.
- Uniformity between medical and allied health staff on use of terminology and classifications of cerebral palsy.
- Evolution and early indicators in very young infants' pre-goal directed movement age. Any particular indicators on GMs for example?
- I think determining the impact once dyskinesia is identified will help raise the importance. What benefits are there to the child and family if we improve identification and measurement? Is it just a label or will it lead to a change in function and participation for the child?
- Assessment of dyskinesia needs to be complemented with functional outcome assessment and individualised goals.
- I would like more information on when DBS is appropriate, including ages, level of involvement, contraindications, long term outcomes.
- Sounds like a great project! Identifying dyskenesias is the first (and probably the easiest) step. PTs having an understanding of the differences in both medical management and best practices for physical intervention when comparing spasticity to dystonia or choreoathetosis is something that should parallel this identification process.
- Perhaps and app for families to use to capture movement patterns they see or that are problematic to daily life to help clinicians understand what is happening and what interventions might be appropriate.
- Only provide an assessment service to children with cerebral palsy, one day a week. Do not treat, and most will have spasticity, with fewer movement disorders.

ABBREVIATIONS

AACPDM: American Academy for Cerebral Palsy and Developmental Medicine

AROM: Active Range of Motion

ASAS: Australian Spasticity Assessment Scale

AusACPDM: Australasian Academy of Cerebral Palsy and Developmental Medicine

Ax: Assessment

BADS: Barry Albright Dystonia Scale

BFMDRS: Burke-Fahn-Marsden Dystonia Rating Scale

CFCS: Communication Function Classification Scale

CPA: Cerebral Palsy Alliance

DBS: Deep Brain Stimulation

DIS: Dyskinesia Impairment Scale

EACD: European Academy of Childhood Disability

GDS: Global Dystonia Scale

GMFCS: Gross Motor Function Classification Scale

HAT: Hypertonia Assessment Tool

ITB: Intrathecal Baclofen

MACS: Manual Ability Classification Scale

MAS: Modifies Ashworth Scale

MD: Movement Disorder

MD-CRS: Movement Disorder – Childhood Rating Scale

MD-CRS (0-3): Movement Disorder – Childhood Rating Scale (0 to 3 years)

MDT: Multidisciplinary Team

MTS: Modified Tardieu Scale

NDT: Neurodevelopmental Therapy

SCPE: Surveillance of Cerebral Palsy in Europe

UDRS: Unified Dystonia Rating Scale

12. APPENDIX 3

SURVEY: ASSESSING DYSKINETIC CEREBRAL PALSY ACROSS THE DOMAINS OF THE ICF

This survey was distributed during a presentation on outcome measures at a CRE-CP Dyskinesia Symposium on 16 March 2017 in Melbourne, Australia.

The purpose was to survey the attendees of the symposium regarding:

- the importance of including outcome measures in the areas of goal setting, quality of life, pain, gross and fine motor
- additional outcome measures that could be utilised to assess children with dyskinetic cerebral palsy to compliment the currently available dystonia and dyskinesia scales and cerebral palsy classifications

In each area, the most commonly applied assessment tools were included for either ranking in importance or selection for inclusion without ranking their importance. If participants were unfamiliar with the individual tools they were given the option to leave selection of individual tools blank.

PARTICIPANTS

Participants (n=113) comprised of 63% physiotherapists (n=70), 13.5% occupational therapists (n=15), 18% medical doctors including rehabilitation specialists, paediatricians, orthopaedic surgeons and neurologists (n=20), one speech pathologist and five participants who did not specify a profession. Two surveys were not completed. All survey results reported are for completed surveys (n=111).

Table 1: Importance of assessment area for inclusion in toolkit

ASSESSMENT AREA	UNIMPORTANT N (%)	SOMEWHAT IMPORTANT N (%)	NEITHER IMPORTANT OR UNIMPORTANT N (%)	SOMEWHAT IMPORTANT N (%)	VERY IMPORTANT N (%)
Goal setting				3 (2.7)	108 (97.3)
QOL/caregiver burden				9 (8.1)	102 (91.9)
Pain				9 (8.1)	102 (91.9)
Gross motor		1 (0.9)	1 (0.9)	19 (17.1)	90 (81.1)
Fine motor		1 (0.9)	1 (0.9)	16 (14.5)	90 (81.1)

GOAL SETTING

Goal setting was overwhelmingly rated as 'very important' to include when assessing children with dyskinetic cerebral palsy (97% of participants, n=108). The two most commonly applied goal setting tools, the Canadian Occupational Performance Measure (COPM) and Goal Attainment Scaling (GAS) were included in the survey for ranking or comment.

The COPM was ranked as the most important and useful tool to include (67.6% of participants) and GAS as the second most important tool to include (34.2%). No additional goal setting tools were suggested by the participants.

Table 2: Goal setting survey results

GOAL SETTING TOOL	RANK 1 N (%)	RANK 2 N (%)	YES – INCLUDE (NOT RANKED) N (%)	NO COMMENT N (%)
COPM	75 (67.6)	12 (10.5)	8 (7.2)	6 (5.4)
GAS	38 (34.2)	48 (43.2)	4 (3.6)	11 (9.9)

QUALITY OF LIFE AND CAREGIVER BURDEN

The assessment of quality of life and/or caregiver burden were also overwhelmingly rated as 'very important' to include when assessing children with dyskinetic cerebral palsy (92% of participants, n=102). A total of five quality of life/caregiver burden assessment tools were included in the survey. These have all either been developed specifically for children with cerebral palsy or had modules/versions developed for use with children with cerebral palsy. This selection was informed by the systematic review of the psychometric properties of Quality of Life measures for school aged children with cerebral palsy (Carlton et al, 2010). The tools include: DISABKIDS – CP Module; Pediatric Quality of Life Inventory CP Module (PedsQL 3.0 CP); Caregiver Priorities and Child Health Index of Disabilities (CPCHILD); Care and Comfort Hypertonia Questionnaire (CCHQ) and the Cerebral Palsy Quality of Life – Child (CP QOL-Child).

The CP QOL and CPCHILD were rated as the most useful and important tools to include in the toolkit with 62% and 48% of participants either ranking them as important or selecting them for inclusion without a ranking. This reflects their specific development for children with cerebral palsy, their robust psychometrics and strong clinical utility. Two participants recommended alternative assessments, neither of which have been utilised with children with cerebral palsy.

Table 3: Quality of life/caregiver burden assessment survey results

	INCLUDE N (%)	RANKED N	YES – INCLUDE (NOT RANKED) N	NO COMMENT N (%)
DISABKIDS CP Module	13 (11.7)	1=0, 2=2, 3=1, 4=2, 5=5	2	98 (88.3)
PedsQL 3.0 CP	33 (27)	1=2, 2=9, 3=4, 4=6, 5=0	12	78 (70.2)
CPCHILD	54 (48.6)	1=18, 2=12, 3=2, 4=1, 5=0	21	57 (51.3)
CCHQ	30 (27)	1=6, 2=6, 3=5, 4=1, 5=1	11	81 (72.9)
CP QOL - Child	69 (62.1)	1=20, 2=9, 3=5, 4=0, 5=1	34	42 (37.8)

PAIN ASSESSMENT

Pain assessment was also considered an important area to assess in children with dyskinetic cerebral palsy. Participants rated it as 'very important' to include (92% of participants, n=102) and 'somewhat important' to include (n=9, 8%). There are four pain assessments that have been validated for use with children with cerebral palsy and found to have good clinical utility for this diagnostic group (Kingsnorth et al 2015). The pain assessment tools selected included: Noncommunicating Children's Pain Checklist – revised (NCCPC-R); Pediatric Pain Interference Scale (PPIS); Pediatric Pain Profile (PPP) and Pediatric Pain Questionnaire (PPQ).

The PPP and PPQ were the two most selected pain assessment tools (n=47, 42.3% and n=42, 37.8% respectively) for inclusion in the toolkit, both of which have strong psychometric data and moderate clinical utility to support their use in cerebral palsy. The only alternative pain assessment suggested was a pain faces visual scale such as the Wong-Baker Faces Pain Scale.

Table 4: Pain assessment survey results

PAIN TOOL	INCLUDE N (%)	RANKED N	YES – INCLUDE (NOT RANKED) N (%)	NO COMMENT N (%)
NCCPC-R	27 (24.3)	1=4, 2=6, 3=0, 4=4	13 (11.7)	84 (75.6)
PPIS	33 (29.7)	1=3, 2=5, 3=4, 4=3	18 (16.2)	78 (70.2)
PPP	47 (42.3)	1=13, 2=5, 3=2, 4=1	26 (23.4)	64 (57.6)
PPQ	42 (37.8)	1=11, 2=7, 3=6, 4=0	18 (16.2)	69 (62.1)

GROSS MOTOR AND FINE MOTOR ASSESSMENT

The assessment of gross motor and fine motor outcomes in children with dyskinetic cerebral palsy was considered 'very important' (n=90, 81%) or 'somewhat important' (n=19, 17% for gross motor and n=16, 14.5% for fine motor) by the participants. Two participants felt the assessment of gross and fine motor outcomes were either somewhat unimportant or neither important nor unimportant (<1%). Selection of gross and fine motor outcomes was based on recommendations by senior physiotherapists or occupational therapists considered experts in their respective fields of clinical practice and on assessments known to have been developed for or validated for use with children with cerebral palsy.

The gross motor outcomes included were: The Gross Motor Function Measure (GMFM); Timed Up and Go (TUG); Walk Tests including 1, 2 and 6 minutes and the Gillette Mobility Scale. The fine motor outcomes included: the Quality of Upper Extremity Skills Test (QUEST); Assisting Hand Assessment (AHA); Melbourne Assessment 2 (Melb 2); Children's Hand-Use Experience Questionnaire (CHEQ); ABILHAND-Kids; Shriners Hospital Upper Extremity Evaluation (SHUEE); Jebson-Taylor Hand Function Test (JTHFT) and Box and Blocks Test (B&B). Participants selected tools rather than ranked them for these two domains of assessments.

Table 5: Gross and fine motor assessment survey results

GROSS MOTOR	GMFM	TUG	WALK TESTS	GILLETTE				
N (%)	84 (75.6)	53 (47.7)	59 (53.1)	34 (30.6)				
FINE MOTOR	**QUEST**	**AHA**	**MELB 2**	**CHEQ**	**ABILHAND-KIDS**	**SHUEE**	**JTHFT**	**B&B**
N (%)	35 (31.5)	47 (42.3)	28 (25.2)	15 (13.5)	15 (13.5)	5 (4.5)	4 (3.6)	7 (6.3)

Additional gross motor assessments suggested included: Quality FM (n=10); Challenge Module (n=4); HiMAT and Berg Balance (n=3 each); 10m walk test (n=2); Chailey (n=1) and use of video for gait and mobility analysis (n=4). Additional fine motor assessments suggested included: AMPS (n=1), task analysis (n=1), BOHA (n=1) and video analysis (n=1).

OTHER TOOLS THAT MAY BE SUITABLE FOR CHILDREN WITH DYSKINETIC CEREBRAL PALSY

There are a variety of other assessment tools that have been developed for or validated with children with cerebral palsy or reported in the literature in studies including children with cerebral palsy. Participants were asked to select those they felt should be included in the toolkit as well as recommend others that could be considered for inclusion. The tools selected in this section included: The Pediatric Evaluation of Disability Inventory (PEDI & PEDI-CAT); Functional Independence Measure for Children (Wee-FIM); Drooling Impact Scale; Viking Speech Scale and Fatigue Scale from the PedsQL.

Table 6: Additional assessment tools survey results

ASSESSMENT DOMAIN	TOOL	N
Function/ADLs	PEDI / Pedi CAT	55
	WeeFIM	37
	Participation	2
Oral facial function	Drooling Impact Scale	66
	Eating/swallow/drinking	10
Fatigue	PedsQL Fatigue Scale	67
Sleep	e.g. Bruni's Sleep Disturbance Scale	36
Communication	Viking	25
	Other communication tool	21
Body functions	Height and weight	7
	Fitness	1
	Vision	1
Parent wellbeing		4
Child mental health	Anxiety (e.g. SCARED, Spence)	1
	Mood	2
Cognition		1

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