Harnessing cognitive strategy use for functional problems and proposed underlying mechanisms in childhood-onset dystonia

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ABSTRACT

Background: There is a significant gap in knowledge about rehabilitation techniques and strategies that can help children and young people with hyperkinetic movement disorders (HMD) including dystonia to successfully perform daily activities and improve overall participation. A promising approach to support skill acquisition is the Cognitive Orientation to daily Occupational Performance (CO-OP) intervention. CO-OP uses cognitive strategies to help patients generate their own solutions to overcome self-identified problems encountered in everyday living.
Purpose: 1. To identify and categorize strategies used by children with HMD to support skill acquisition during CO-OP; 2. To review the possible underlying mechanisms that might contribute to the cognitive strategies, in order to facilitate further studies for developing focused rehabilitation approaches.
Methods: A secondary analysis was performed on video-recorded data from a previous study exploring the efficacy of CO-OP for childhood onset HMD, in which CO-OP therapy sessions were delivered by a single occupational therapist. For the purpose of this study, we reviewed a total of 40 randomly selected hours of video footage of CO-OP sessions delivered to six participants (age 6–19 years) over ten intervention sessions. An observational recording sheet was applied to identify systematically the participants' or therapist's verbalizations of cognitive strategies during the therapy. The strategies were classified into six categories in line with published literature.
Results: Strategies used by HMD participants included distraction, externally focussed attention, internally focussed attention, emotion self-regulation, motor imagery and mental self-guidance. We postulate different underlying working mechanisms for these strategies, which have implications for the therapeutic management of children and young people with HMD including dystonia.
Conclusions: Cognitive strategy training can fundamentally change and improve motor performance. On-going work will address both the underlying neural mechanisms of therapeutic change and the mediators and moderators that influence how change unfolds.

1. Background

Childhood-onset hyperkinetic movement disorders (HMD), including dystonia, are a heterogeneous group of disorders with varied aetiologies, motor severity, age at onset and impact of their movement disorders in daily life [1]. Dystonia is the most prevalent movement disorder phenotype and dyskinetic cerebral palsy (CP) the commonest aetiological sub-group of disorders within HMD. Dystonia has been defined as a HMD characterized by involuntary, sustained, or intermittent muscle contractions that cause twisting and repetitive movements, abnormal postures, or both [2]. Several underlying pathophysiological mechanisms have been identified, including reduced inhibition

throughout the nervous system [3,4], exaggerated cortical plasticity [5], abnormal processing of sensory information [6–8] and pathologically enhanced low frequency neuronal oscillations [9,10] which leads to increased variability in motor performance [11–13]. These physiological abnormalities arise from dysfunctions within the basal ganglia, cortex, cerebellum or their inter-connections as part of the sensorimotor network [14–17].

According to Sival et al., 2021, the cerebellum plays a 'pivotal role' by enabling information processing and modulation of timing, direction and fluency of co-ordinated motor performances. Healthy and typically developing children may exhibit physiological, i.e., immature movement patterns that might wrongly be described as ataxic or dystonic [18, 19]. This is consistent with a view that all infant movements are initially involuntary and show features consistent with physiological dystonia [20] that gradually diminish as surround inhibition is more successfully deployed over time during specific learned actions.

Two recent international consensus papers, have agreed on key areas of future research to improve understanding of the causal role the cerebellum might have in dystonia from compensatory and contributory effects [21] and efforts towards a system-level view of cerebellar functional interplay with basal ganglia and cortical structures [22]. This understanding of the cerebellum acting as a potential node in dystonia might help with treatment modalities in dystonia with some authors proposing treatments around motor learning using sequence-based learning [23].

Anticipatory control appears a fundamentally early-acquired strategy for reaching a moving target, depending on interpretation of object spatial localisation, speed and direction of travel of and calculation the timing of initiation and speed of a reaching action likely to succeed in grasping the object.

This anticipatory control is already well developed in the healthy one year old but disrupted in the infant with perinatal antecedents and early cerebral palsy. Infants are able to calculate the time required to reach a moving object, but this diminishes as an object's speed of travel increases. Healthy infants compensate for this by starting to reach earlier, but infants with early signs of HMD are unable to start reaching earlier which results in missing the target [24]. A failure to develop such anticipatory control may result in unsuccessful motor task achievement and a failure of developing automaticity or *habitualisation* of action.

The basal ganglia, a group of subcortically interconnected nuclei, have been established as fundamental for the performance of habitual (automatic) and goal-directed movements [25]. The ability to 'habitualise' actions after a period of motor learning is essential for smooth, effortless action which nevertheless must at all times be capable of being over-ridden by goal-directed corrective action [25,26]. Habitualisation of mal-adaptive motor strategies actions may simply ingrain failure while also inhibiting internalisation of successful or useful motor habits. Early onset dyskinetic motor behaviours thus archive poor motor strategies, reinforcing the maladaptive networks.

In childhood-onset HMD, a systematic review of medical and surgical interventions for dyskinetic CP are both scarce and of weak efficacy [27] including pharmacological management [28,29]. Research outlining best-practice for rehabilitation in childhood-onset HMD including dystonia or dyskinetic CP is lacking even if such recommendations include occupational and physical therapists in providing therapy by mobilizing joints, limiting contractures, establishing exercise programs, and providing assistive devices [30].

To improve motor performance in the individual's self-selected goals, the efficacy of the Cognitive Orientation to daily Occupational Performance (CO-OP) Approach [31] was explored by Gimeno and colleagues (2019) and yielded positive results pertaining to skill acquisition and performance in children with HMD who had undergone pallidal Deep Brain Stimulation (DBS) [32,33]. CO-OP is a client-centred, performance-based, problem-solving approach that enables skill acquisition through a process of cognitive strategy use and guided discovery [31].

Research on the efficacy of CO-OP and the use of cognitive strategies with regards to achieving motor-based performance goals has been conducted in various populations including children and adults with other neurological disorders [34]. Previous research with these populations has yielded promising results for the use of CO-OP in regards to maintenance of skill acquisition, generalization of learning, and transfer of learning to similar tasks [35].

Similar types of performance problems tend to require kindred types of cognitive strategies, allowing them to be grouped into classes. In the early years of research on the CO-OP approach, cognitive strategies were identified and categorized from studies using the intervention with individuals with Developmental Co-ordination Disorder (DCD) [36]. While these have continued to prove useful in studying the efficacy of the CO-OP approach in some other motor disorders groups, it is likely that those with needs quite distinct from children with DCD might need different strategies. Children and young people with HMD may require unique strategies to meet their distinct performance demands. Analysing characteristics of these strategies could contribute to further understanding and elucidation of underlying mechanisms to develop personalised rehabilitation.

This study aims to (i) investigate the cognitive strategies individuals with childhood-onset HMD use to support skill acquisition and performance during CO-OP sessions; and (ii) postulate potential underlying mechanisms for further research when developing rehabilitation interventions.

2. Method

2.1. Study design

Single group prospective observational study, with secondary analysis of previously collected video-recorded data from a single case experimental design study exploring the efficacy of the CO-OP Approach for childhood-onset HMD in individuals who have undergone pallidal DBS [32]. The protocol for the main study is available elsewhere [37].

Behavioural observation was utilized to identify strategies used by participants during CO-OP intervention sessions. Ethical approval for this secondary study was obtained from the ethics committee at the University of Toronto. The primary study was approved by the NHS Health Research Authority Oxford A Research Ethics Committee (14/SC/1159) and was registered with the ISRCTN Registry (ISRCTN57997252).

2.2. Participants

From the nine available participants in the single case experimental design study [32] six children and young people (3 male, 3 female) had completed video-recorded data sets for all goals and all therapy sessions. The three children not included in the secondary data analysis had videos of goals such as swimming or bike riding where sound and video were not of enough quality for data extraction to take place. Participants had a diagnosis of childhood-onset HMD and DBS in situ. Age range was 9–19 years. Gross Motor Function Classification System (GMFCS) [38] and Manual Ability Classification System (MACS) [39] levels, and cognitive ability/executive function using the Behavior Reported Inventory of Executive Function (BRIEF) [40] are shown in Table 1. Whilst the MACS and GMFCS levels were validated for individuals with cerebral palsy, the use of this classification for other movement disorders has been used before as GMFCS/MACS-equivalent [41–43]. Inclusion criteria were a diagnosis of childhood-onset HMD other than neurodegenerative conditions, age 6–21 years, and having pallidal DBS in situ with no signs of infection. Participants were also required to express a willingness to participate in CO-OP sessions, require adult assistance to complete age appropriate tasks, and have sufficient receptive and expressive communication to follow simple instructions in English. Participants were excluded if they presented with pure spasticity or a

Table 1
Participant Demographics.

Participant #	Age (years)	Diagnosis	Cognitive impairment	BRIEF (T score) BRI | MI		GMFCS/MACS Level	Self-Selected Treatment Goals
1	9y 8 m	Idiopathic Dystonia	No	86	64	GMFCS I/MACS II (equivalent)	1. Catching a tennis ball 2. Making a drink 3. Doing a tie
2	17y 4 m	Dyskinetic CP - hemidystonia	Yes	67	59	GMFCS I/MACS II	1. Doing a ponytail 2. Painting nails 3. Doing up shoelaces
3	17y 6 m	Dyskinetic CP - HIE	No	62	57	GMFCS II/MACS III	1. Opening a yoghurt 2. Carrying a bowl of cereal 3. Achieving neat handwriting
4	18y 11 m	Inherited Dystonia – Benign Hereditary Chorea	No	64	52	GMFCS I/MACS II (equivalent)	1. Carrying a hot drink and a plate 2. Turning bacon in the grill and making a sandwich 3. Applying eye liner
5	13y 10 m	Dyskinetic CP - Kernicterus	No	46	42	GMFCS II/MACS IV	1. Brushing teeth 2. Putting T-shirt on 3. Making a hazelnut spread sandwich
6	14	Idiopathic Dystonia	No	62	60	GMFCS I/MACS III (equivalent)	1. Carrying a glass of water 2. Cutting with knife and fork 3. Doing up shoelaces

Abbreviations: BRIEF: Behavior Rating Inventory of Executive Function; BRI: Behavior Regulation Index' MI: Metacognition Index; GMFCS: Gross Motor Function Classification System; MACS: Manual Ability Classification System; CP: Cerebral Palsy; HIE: Hypoxic Ischaemic Encephalopathy.

mixed phenotype where spasticity was dominant, dystonia arising due to a neurodegenerative condition, signs of DBS infection, or scheduled surgical treatment during the study period.

2.3. Data collection

The video data used for this study consisted of approximately 2400 min (40 h) video recorded CO-OP intervention sessions out of a total of 60 h of therapy intervention sessions available for the 6 participants. The videos observed, were randomly selected from ten intervention sessions (66% of available data), ranging from 45 to 60 min in length, for each of the six participants.

The videos were studied in a staged manner. Initially, purposive sampling of ten CO-OP sessions for three participants were viewed in their entirety. This included a total of over 1800 min (30 h) of video recorded data. Next, to ensure data saturation was achieved, three sessions were randomly selected from the treatment videos by an independent party for the remaining three participants (i.e., one CO-OP treatment session within sessions one to three, four to six, and seven to ten). An online randomization resource was used to select five different minute-marks in a session in which to observe 2-min of video.

An observational recording sheet was developed for the purpose of systematically identifying the participant's or therapist's verbalizations of cognitive strategies during the CO-OP intervention sessions. The recording form incorporated the global task, the participant's goal, and dialogue of the therapist or participant when verbalizing a strategy.

The criterion for classifying a strategy included: (1) strategies discussed in words either by the therapist or participant; (2) verbalization of a strategy either immediately before or after it was performed resulting in change of an observable motor behaviour. Whenever the researchers identified the successful implementation of a verbalized strategy, it was immediately recorded on the observational recording form. No attempts to calculate frequency of strategy use was conducted *as researchers were interested in identifying novel strategies rather than their frequency.*

Inter-observer reliability was established between the two researchers (KB & MM) by independently reviewing and identifying strategies with transcripts of all ten CO-OP treatment sessions for one participant (participant 6). After discussion and consensus building around observations, researchers were in a hundred percent agreement in identifying all individual specific cognitive strategies observed within

each CO-OP session. Researchers then independently viewed the remaining video recorded data and identified verbalized strategies.

2.4. Data analysis

After viewing the first three participants' ten CO-OP sessions (participants 3,4, and 6) and identifying all strategies, the two researchers collaboratively began classifying strategies using definitions of cognitive strategies from literature pertaining to the CO-OP Approach on strategy-use with children with DCD [44].

Strategies that did not fit the observed behaviour or definitions from past CO-OP DCD studies were identified as novel strategies. Researchers then proceeded to code novel strategies. Categories were developed to group novel strategies with similar observed behaviour and purpose when performing a task. Researchers (MM, KB) consulted with senior authors (HP, HG) to verify the coding of strategies.

3. Results

Behavioural observation of participants during intervention sessions identified a number of strategies used. Many were similar to those described for the DCD population such as body positioning, attention to task, or task specification [44], while others were novel cognitive strategies, not previously reported in CO-OP literature, identified for childhood-onset HMD including dystonia. These novel strategies were themed under six main types following several face-to-face discussions and teleconferences to reach consensus among all the authors. The observed behaviour and examples of verbalizations of the strategy made by the participant or therapist for each novel cognitive strategy are defined in Table 2. The mechanisms underlying these strategies are postulated in the discussion section.

4. Discussion

There is a significant knowledge gap around rehabilitation techniques and strategies that can help children and young people with HMD including dystonia. Contrary to the literature available on strategies for other movement disorders such as Parkinson's Disease [45], a systematic evaluation of what works, how it works and for whom, is not available. Current advice is therefore based largely on expert opinion [30,46] and not on research.

Table 2
Novel Strategies Observed during CO-OP Treatment Sessions.

Novel Strategy	Observed Behaviour	Example of Verbalization
Distraction (Passive, Active, Increasing cognitive load)	Something that draws the client's attention away from focusing on the task, or the bodily movements associated with the task e.g., something that encourages a client to direct less attention to the task or components of the task.	*Passive*: "Having TV or music playing in the background" *Active*: "I was talking to my dad" *Increasing cognitive load:* "Do some complex math in your head, you know like 100 minus 7″
Emotion Regulation	A reminder to elicit behaviours or visualizations that reduce anxiety or tension, either prior to or during task performance.	"Breathing, let's think about the breathing" "Thinking of Christmas or happy thoughts"
Motor Imagery	The individual imagines and rehearses in their mind the movement required to perform the task.	"Think of the pulling movement before you do it"
Mental self-guidance	Any verbalization for the formation of mental images or likenesses of people/ places/things that support task performance.	"I am imagining a carpentry level being stuck to the tray"
Externally focused attention	Any verbalization of directing and sustaining attention to an object or location related to the task, outside the body. e.g., the final destination or the object being handled and carried	"I was thinking more about the cup instead of my hand" "As you're doing it, you're thinking of where you're going with it"
Internally focused attention	Any verbalization of directing or sustaining attention to the body, whole or in part, that is actively being utilized during task performance.	"Your plan was looking at the hand"

Our study identified 16 strategies, some already described in CO-OP studies with other populations, and six unique categories of cognitive strategies for childhood-onset HMD, which were generated and used by participants to engage in meaningful activities and support skill acquisition during CO-OP sessions. We postulate that these strategies have different underlying mechanisms, which in turn has implications for therapeutic management in this population. Below we discuss each strategy in the context of known neuroanatomical pathways and neurophysiological mechanisms that are implicated in dystonia and speculate on their potential underlying physiological basis.

Distraction (Passive, Active, Increasing cognitive load): By using distraction (e.g. television or music) whilst carrying out the task/goal being practiced, participants were able to improve their performance and reduce their involuntary movements during the task. The same result was possible when participants were completing goals whilst actively doing another activity such as talking to someone. Some strategies aimed to increase their cognitive load, i.e. the amount of mental effort and working memory required while performing a task [47]. Examples included reciting numbers in another language, or subtracting 7 from 100. One hypothesis for how this strategy works is that increasing cognitive load whilst performing an activity allows participants to ignore or block-out thoughts of previous failed trials with their goal. Stress and anxiety have been shown to reduce motor performance quality [48] by increasing attention to negative thoughts [49], which in the case of individuals with HMD may be related to previous negative experiences of failed motor performance. These negative thoughts may impair working memory [49], hinder motor performance and, in many cases, increase the amplitude and severity of involuntary movements and postures. Thus, in young people with HMD with competing brain programs, a reduction of performance anxiety using distraction

strategies may allow better motor performance. Another potential explanation is the shift in attentional focus towards something unrelated to the task, (e.g. a conversation or the television), allowing dystonia symptoms to remain outside focal awareness and making movements more automatic or habitual, therefore reducing the interoceptive sensory cues and the associative thoughts hindering motor performance [50]. Since attempts at voluntary movements are known to increase the unwanted production of involuntary movements even before the voluntary movement is produced, distraction or shift in attentional focus may block the production of the involuntary movement because the cognitive pathway for intention is 'preoccupied'/in use. This idea is similar to the limited attentional capacity theory in pain where the more attentional resources are used by distraction, the less they are available for perceiving pain [51].

Basal ganglia loops with projections from functional territories of the cerebral cortex (limbic, associative, sensory and motor) highlight the co-adjacency of systems underpinning drive and reward, motor processing and memory storage of the event and motor performance respectively. These systems are essential for performance of habitual and goal-directed movements [25].

Practicing maladaptive motor strategies reinforces maladaptive habitual motor function and persistence of overtly goal-directed actions which are relatively coarse and require intensely conscious (rather than habitual) monitoring [25]. Since the cerebello-thalamic-cortico-basal ganglia loops encompass sensorimotor, associative and limbic functions, unsuccessful strategies and motor goal failure negatively reinforce performance leading to a dwindling motor drive.

Conversely, successful strategies such as analysing the approach to motor task completion promotes a generalized 'can do' approach to familiar and unfamiliar activities. In this sense, CO-OP may be seen as a strategy for generating successful 'habitualisation' of specific motor tasks as well as a technique applicable to future 'habitualisation' demands.

4.1. Emotion regulation

Clinical anxiety is also present in children and young people with dystonia due to injuries/disturbances to the basal ganglia [52,53]. Many of the participants found that when they utilized strategies that directed their thoughts or focus to non-anxiety provoking elements, they were able to perform their chosen goal with greater satisfaction and consistency.

Participants verbalized the strategy of stress-reduction and calming their emotional state before, or during task performance if an error was made. Anxiety has been reported in children and young people with dystonia and poor self-esteem is related to higher levels of anxiety and emotional difficulties [52]. In turn, individuals with anxiety display attentional bias towards potential threatening sources of information and compromise their ability to switch their attention to the actual goal performance [54]. Children and youths with HMD may comprehend what sequence of movements is required; however, the ever-changing state of fluctuating postures and movements, influenced by emotional factors and attempts at volitional movement, triggers further involuntary and undesired movement and postures that interfere with task performance.

We have discussed the positive effect of distraction as a cognitive strategy, but it is important to differentiate this from situations when distraction (understood as the threatening source of information) can challenge our ability to maintain focus on goal-relevant information. We propose that emotionally arousing stimuli (previous failed experiences when attempting these tasks) act as potential distractors, leading children and young people to re-allocate cognitive resources [55]. This emotional distractibility impairs performance and leads to a form of performance anxiety. The strategy of auto-relaxation before or during task performance might in turn reduce the detrimental effects of emotion on cognitive functions and help young people to problem-solve.

The cognitive-affective interaction involves the interplay between the dorsal neural and ventral systems with dysfunction in the prefrontal-striato-limbic-thalamus networks [56]. A proposed mechanism of emotion regulation is the allocation of attention to a different source and therefore this emotion regulation strategy is closely related to the first strategy of distraction. Feeling in control of performance can modulate the effect of stress on cognitive functioning and the associated neural mechanisms [56].

4.2. Motor imagery and mental self-guidance

Some individuals utilized the strategy of motor imagery, in which they imagined and rehearsed in their mind the movement required to perform the task, prior to actually carrying it out. For example, participants would think about the movement required to open a pot of yoghurt or the trajectory of the pencil when putting on eyeliner.

While mental practice required imagining the movement required, the strategy of mental self-guidance required imagining something external to the task that would support performance. This was seen in circumstances where participants carried an object (i.e., a bowl or cup) containing liquid, so they would imagine a carpenter's level to aid holding the bowl level and avoiding spillage.

Motor imagery and action observation (observing someone doing the movement) have been identified as compensatory strategies in Parkinson's disease (PD) and are thought to compensate for reduced automaticity by mental simulation of the action without the execution [45]. The underlying mechanism is thought to relate to activation of the mirror neurone system. Transcranial magnetic stimulation studies show that in healthy subjects, imagery-related mechanisms modulate the excitability of sensorimotor pathways [57–59]. Physiological correlates of motor imagery are abnormal in focal hand dystonia [58], PD [60] and spastic cerebral palsy [61], but motor imagery is indeed used as a compensatory strategy by patients with gait impairment or upper limb freezing in PD [45,62].

Motor imagery is also associated with changes in oscillatory activity in the EEG, namely suppression of mu (8–12Hz) activity over sensorimotor cortex [63,64], although the degree of suppression is less than that seen during movement observation [63], which in turn is less than that seen during active movement [65]. Nevertheless, the suppression of mu activity during motor imagery has been considered to reflect an association with the mirror neurone system.

Suppression or event-related desynchronisation of mu activity in relation to both real or imagined movement is considered to reflect activation of the sensorimotor network [66], so again, this could explain how motor imagery helps as a strategy – i.e., enabling the individual to "practice" activating the relevant sensorimotor circuit for the task. Interestingly, a recent study found that modulation of mu activity in response to a proprioceptive stimulus is reduced in young people with dystonia compared with typically developing children [67].

Motor imagery and rehearsal may provide, therefore, an important strategy for increasing repetition, excitation and neuroplasticity of successful task performance, which in turn may assist with fading the competitive maladaptive plasticity.

4.3. Externally focused attention

Externally focused attention involves participants choosing to focus attention on an object or location external to themselves or the task, directing attention away from their involuntary, tremulous or jerky movements. Within the PD literature, cueing (including external cueing) is reported as one of the main "compensatory" strategies, particularly for gait impairments. It is thought that the involved mechanism introduces a goal-directed behaviour and therefore circumnavigates areas of the basal ganglia involved in automatized motor control [45]. Whilst it is postulated that this can help other basal ganglia areas less severely affected by loss of dopaminergic innervation to become active in this goal-directed behaviour, this hypothesis does not necessarily hold for childhood-onset HMD with no known loss of dopaminergic innervation. Therefore, the neural networks involved for this population are still unknown.

4.4. Internally focused attention

Internally focused attention involves the participant sustaining attention on a body part that is actively participating in the task. The difference between externally and internally focused attention and the strategy of distraction is that participants still attend to aspects of the task they are performing as opposed to using distracting stimuli unrelated to task performance. Internally focused attention, also termed "internal cueing", has been observed in PD [45]. In our study, children and young people often verbalized this strategy, which facilitated its classification.

Several studies have shown that attention can modulate the somatosensory mu rhythm that was described above in relation to mental imagery and sensorimotor processing. For example, Anderson and Ding (2011) demonstrated that lateralised spatial attention reduces mu power over the sensorimotor cortex contralateral to the hand to which the participants were directing their attention [68]. While Woodruff and Klein (2013) found that mu suppression in response to observation of a hand movement task was eliminated when the participant had to simultaneously perform a word generation distraction task [69]. They concluded that simple fixation of the eyes on an action without focusing attention is insufficient to induce mu suppression. Thus, it is possible that internally focussed attention can work as a strategy for some young people with HMD by enhancing sensorimotor processing in a similar way to mental imagery. It could also be envisaged that these two strategies might work in combination.

Findings from a recent study indicate that internally focussed attention can be optimised when integrated with proprioceptive information relevant to the task [70], suggesting that combining this strategy with biofeedback could enhance its effectiveness.

Indeed, a study in children with dystonia found that using visual feedback of the degree of co-contraction during a reaching task allowed the children to exert some control over their co-contraction [71]. Another recent study found that the optimal attentional strategy varies between individuals and their motor imagery ability, suggesting that an individualised approach to selecting strategies for therapy/rehabilitation may be advantageous [72].

4.5. Summary and relevance for clinical practice

The strategies described above may enhance or damp down activity within given functional or dysfunctional neural circuits, shifting the balance towards improved motor control. Since dystonia is a network disorder, with different dystonia sub-types arising from various parts of the sensorimotor network, involving basal ganglia, thalamus, sensorimotor cortex and cerebellum, or their interconnections, it follows that different individuals may find different strategies to be more or less beneficial. Understanding these differences between individuals and the mechanisms underlying the various strategies in the repertoire, could allow us to optimise therapy on a more individual basis.

There is currently a lack of research supporting non-pharmacological or surgical interventions to aid individuals with childhood-onset HMD perform activities they want, need, and are expected to perform, even following DBS. Emerging evidence suggests that the CO-OP Approach may be effective in enabling skill performance and acquisition of children and youths with HMD and DBS during self-selected activities and participation goals [32,33,73]. The current study substantiates this evidence by identifying the novel strategies used by these children, which clinicians may incorporate within their repertoire to assist guided discovery and development of plans through the CO-OP Approach. This research also contributes to the current body of knowledge on

interventions for children with HMD and describes strategies that may be applicable to various adult populations with movement disorders.

4.6. Limitations

A number of limitations to this study must be acknowledged. The small sample size limits how generalizable these strategies are to the population of childhood-onset HMD. Future research with a larger sample size is required to confirm if the novel strategies identified here can be easily implemented and generalized to the wider HMD population. This study aimed to identify which strategies people with HMD used during their CO-OP treatment rather than evaluating how many and which strategies each person used and the relationship with outcome. These are important future research questions.

A single CO-OP trained therapist administered the treatment for all participants, which questions how other therapists may be able to help generate these strategies within the CO-OP Approach.

5. Conclusion

Cognitive strategy training can fundamentally change and improve motor performance. This study identifies novel cognitive strategies used by children with HMD, and considers how they might help improve motor performance within the context of the network model of dystonia. Understanding why treatment approaches such as CO-OP work, what the underlying neural mechanisms are, and how change comes about, are important in order to design successful therapeutic and rehabilitation strategies and to optimise the therapeutic change obtained. Further work on mediators and moderators is necessary to understand how change unfolds.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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