Clinical presentation of spasticity and passive range of motion deviations in dyskinetic cerebral palsy in relation to dystonia, choreoathetosis, and functional classification systems

Objectives: To map the presence, severity, and distribution of spasticity and passive range of motion (pROM) deviations in dyskinetic cerebral palsy (DCP), and to explore their relation with dystonia, choreoathetosis, and functional abilities.

Methods: This cross-sectional study included 53 participants with DCP. Spasticity was assessed with the Modified Ashworth Scale, limited- and increased pROM (hypermobility) with a goniometer, dystonia and choreoathetosis with the Dyskinesia Impairment Scale, gross motor and manual abilities with corresponding functional classification systems.

Results: Spasticity and limited pROM were correlated with dystonia of the upper limbs ($0.41<r_s<0.47$, $<0.001<p<0.002$) and lower limbs ($0.31<r_s<0.41$, $0.002<p<0.025$), and both functional systems of gross motor ($0.32<r_s<0.51$, $<0.001<p<0.018$) and fine manual abilities ($0.34<r_s<0.44$, $0.001<p<0.014$). Hypermobility is correlated only with choreoathetosis of the lower limbs (0.44, p=0.001).

Conclusions: Coexisting spasticity and pROM deviations in DCP are functionally limiting and should be addressed accordingly. Hypermobility may lead to an increased luxation risk.

Keywords: cerebral palsy, spasticity, dystonia, choreoathetosis, passive range of motion, functional classification system

Introduction

Cerebral palsy (CP) comprises a group of developmental disorders of movement and posture, classified based on the dominant movement disorder such as spasticity, dystonia, choreoathetosis, and ataxia.[1] Accompanying impairments, including later-developing musculoskeletal problems, non-motor neurodevelopmental problems and/or sensory problems are often present.[1]

Dyskinetic CP (DCP) is the second most common type of CP after spastic forms with a prevalence up to 15%.[2] DCP is defined by abnormal postures or movements due to impaired muscle tone regulation and impaired movement control and coordination.[2] In DCP, dystonia and choreoathetosis are the dominant movement disorders, often simultaneously present.[2] Dystonia is characterized by involuntary movements and abnormal postures due to sustained muscle contractions, whereas choreoathetosis is characterized by rapid, continuously changing hyperkinetic movements.[2-4] The severity of dystonia is higher than the severity of choreoathetosis with larger negative impact on functional motor abilities.[5,6] As a result, treatment options are strongly focused on dystonia as its impact on daily life activities, societal participation, and quality of life is larger.[7]

Beside the predominant dystonia and choreoathetosis, the coexistence of spasticity as a concurrent hypertonia component (i.e. mixed hypertonia) in DCP is often observed.[8-10] Previous studies report high percentages of coexisting spasticity, such as 61% and 71% of the participants with DCP.[11,12] This is clinically relevant as different movement disorders like spasticity and dystonia require a different treatment management, thus unidentified coexisting movement disorders may impact the success of an outcome following an intervention.[8-10] For instance, in spastic CP, a selective dorsal rhizotomy or tendon transfer surgery may result unsuccessful due to unrecognized coexistence of dystonia.[8,13,14] In DCP, the implications that

spasticity may have as a coexisting movement disorder are not fully explored, but it is likely to impact clinical outcomes if left unidentified and untreated. Previous research does not report the severity and the distribution of spasticity in different body regions in the DCP population. This knowledge gap makes the management of DCP even more challenging.

Apart from the coexisting movement disorders, CP is often characterized by accompanying musculoskeletal abnormalities resulting from the combination of muscle weakness and hypertonia.[15] Individuals with DCP are at high risk of developing musculoskeletal deformities and their clinical implications are only scarcely investigated.[16] Passive range of motion (pROM) is widely used to inform clinical decisions on the therapeutic management of CP, especially important in preventing the development of contractures.[17] pROM of individuals with CP is lower than their typically developing peers, further decreasing with an increase in age and functional limitations.[16-19] The number of participants with DCP included in these studies is low compared to spastic CP, thus conclusions cannot be inferred merely for the DCP population. In addition, clinical practice shows that beside limited pROM, joint hypermobility (i.e. increased pROM)[20] is present in individuals with DCP, often linked to increased luxation risk and pain. To the best of researchers' knowledge, joint hypermobility has not been described before in individuals with DCP.

Identifying all coexisting movement disorders and impairments during assessment of patients with DCP may thereby hold important clinical information which would assist tailoring an appropriate management strategy leading to improved clinical outcomes.[2,8-10,12,13]

Therefore, this retrospective study aims to increase insights in the clinical presentation of spasticity and pROM deviations in individuals with DCP, and to assess the relationships of spasticity and pROM deviations with dystonia, choreoathetosis, and functional motor abilities.

Materials and Methods

Participants

In this retrospective cross-sectional study, participants, aged 6 to 22 years were recruited from five Flemish special education schools for motor disabilities. Inclusion criteria were (1) diagnosed with DCP by a paediatric neurologist and (2) a good ability to understand and follow instructions. Exclusion criteria were (1) change in medication within the previous 3 months and (2) orthopaedic or neurosurgical interventions within the previous 12 months. Ethical approval was obtained from the Medical Ethics Committee UZ KU Leuven. Assent to participate was obtained by all participants and informed consent forms were signed by them or their parents.

Assessment and procedure

Clinical examinations to assess the presence and severity of spasticity, pROM, dystonia, and choreoathetosis were administered by an experienced physiotherapist at the special education schools of the participants. Spasticity and pROM deviations were each measured twice on the same occasion, and the score representing the higher impairment was used for further analyses. Functional motor abilities (i.e. gross motor and manual abilities) were evaluated the same day as the other clinical examinations.

Spasticity was measured using the Modified Ashworth Scale (MAS).[21] The MAS describes the resistance of a muscle during a passive stretch on a six-point ordinal scale (scores 0, 1, 1.5, 2, 3, and 4 where a score of 0 is assigned if the muscle tone is normal, and a score of 4 is assigned if no motion is possible due to rigidity). The MAS in the current study was measured in 22 muscle groups, that is, 11 muscle groups in each side of the body. In the upper limbs (UL), spasticity was measured for the elbow flexors and extensors, for the forearm supinators and pronators, and for the wrist dorsiflexors and palmarflexors. In the lower limbs (LL), spasticity

was measured for the hip abductors (0 and 90 degrees), for the hamstrings, for the soleus, and for the gastrocnemius.

pROM was determined using goniometric measurements[22,23] following a standardized protocol used in the Movement Lab of the University Hospital Leuven. pROM in the current study was calculated in 38 joint movements, that is, 19 joint movements in each side of the body. In the UL, pROM was measured in a supine position for the shoulder (flexion, extension, abduction, adduction) and for the elbow (extension, flexion), and in a supine position with a 90° elbow flexion for the wrist (dorsiflexion, palmar flexion). In the LL, pROM was measured in a supine position for hip flexion (Thomas test), in a supine position with extended knees for hip abduction and hip adduction, in a supine position with flexed knees for hip external- and internal rotation, and in a prone position for hip extension. Knee flexion and knee extension were assessed in a supine position. Ankle dorsiflexion was assessed using the Silfverskiöld test[24] with knees both flexed and extended to differentiate between gastrocnemius and soleus muscle contraction. Ankle plantarflexion was measured in a supine position with knees flexed.

Presence and severity of dystonia and choreoathetosis were assessed using the Dyskinesia Impairment Scale (DIS).[25] The DIS has a subscale for dystonia (DIS-D) and choreoathetosis (DIS-CA), measuring duration (i.e. the amount of time that dystonia and choreoathetosis were present, range 0-4) and amplitude (i.e. the range of motion in which dystonia and choreoathetosis are present, range 0-4). In the current study, the DIS was assessed in the UL (proximal and distal) and the LL (proximal and distal).

The Gross Motor Function Classification System (GMFCS)[26,27] and the Manual Ability Classification System (MACS)[27,28] were used to assess the gross motor and fine manual abilities of the participants.

Outcome measures

Presence of spasticity was calculated by dichotomizing the MAS data per muscle group.[29,30] A 0-score was assigned when spasticity was absent in a muscle group (MAS score = 0) and a 1-score when there was a sign of spasticity present (MAS score = 1, 1.5, 2, 3, 4). Presence of spasticity for the whole body, for the UL, and the LL was calculated by summing the dichotomised scores per participant per muscle group, accounting for both the right and left side of the body. This leads to a possible summed score from 0-22 for the whole body, 0-12 for the UL (0-4 for the UL proximal, 0-8 for the UL distal), and 0-10 for the LL (0-6 for the LL proximal and 0-4 for the LL distal). The summed score was converted into a percentage relative to the maximum score per body segment. A median percentage score and an interquartile range was then calculated for the sample. Additionally, presence of spasticity for each measured muscle group was calculated among the sample. This has been done by scoring each measured muscle group of every participant from 0-2 (0 = no spasticity, 1 = spasticity only present in left or right side of body, 2 = spasticity present in both sides of the body), and subsequently summing the scores of all participants for that particular muscle group. This summed score was then converted into a percentage relative to the maximum possible score (Maximum possible score is 106, namely, 53 (participants) * 2 (score if presence in both left and right side of body).

Severity of spasticity per body segment was calculated by summing the MAS scores (0, 1, 1.5, 2, 3, 4) of all measured muscle groups per participant (22 muscle groups),[31,32] and converting this score into a percentage relative to the maximum possible score of a body segment, i.e. 48 for the UL (16 for the UL proximal and 32 for the UL distal), and 40 for the LL (24 for the LL proximal and 16 for the LL distal). A median percentage score and an interquartile range was then calculated for the sample. Severity of spasticity for each muscle group was calculated among the sample by summing the MAS scores of all participants for the left and right side of each muscle group. This summed score was then converted into a percentage relative to the

maximum score of 424, that is, 53 (participants) * 2 (body sides) * 4 (maximum possible MAS score) = 424. The mode of spasticity severity aimed to inform which MAS score reoccurred most often in each muscle group, and it was calculated excluding the 0 scores (i.e. including only MAS scores 1, 1.5, 2, 3, and 4).

pROM was calculated for both limited pROM (pROM-) and increased pROM (pROM+ i.e. joint hypermobility) relative to the normal ROM. First, the measured ROM per participant for each joint movement was subtracted from the respective normal ROM, and the obtained value was either negative (indicating pROM-) or positive (indicating pROM+). For instance, for ankle dorsiflexion where 20° is considered as normal ROM, the calculation would be as follows: 10° (measured pROM) – 20° (normal ROM) = -10° (indicating pROM-); or 50° (measured pROM) – 20° (normal ROM) = 30° (indicating pROM+). Second, a 25^{th} (first quartile Q1) and 75^{th} (third quartile Q3) percentile was calculated separately for the negative values (i.e. pROM-) and for the positive values (i.e. pROM+).[32] All pROM measurements were transformed to a 3-point ordinal scale ranging from 0 to 2, using their 25^{th} and 75^{th} percentile as cut-off values. As such, for pROM-, 0 was assigned for values higher than the 75^{th} percentile, indicating no or slight limited pROM; 1 was assigned for values between the 25^{th} and the 75^{th} percentile, indicating moderately limited pROM; and 2 was assigned for values lower than the 25^{th} percentile, indicating severely limited pROM. For pROM+, 0 was assigned for values lower than the 25^{th} percentile, indicating no or slight increase in pROM; 1 was assigned for values between the 25^{th} and the 75^{th} percentile, indicating moderately increased pROM; and 2 was assigned for values higher than the 75^{th} percentile, indicating severely increased pROM. These assigned ordinal scores were summed up for pROM+ and pROM- separately, and converted into a percentage score relative to the maximum possible score, i.e. maximum possible score of 36 for the UL (28 for the UL proximal and 8 for the UL distal), and 40 for the LL (32 for the LL proximal and 8 for the LL distal). A median percentage score and an

interquartile range was then calculated for the sample. Additionally, pROM- and pROM+ for each joint movement separately were calculated among the sample by summing the ordinal scores of all participants for each joint movement, and converting the summed score into a percentage relative to the maximum possible score of 212, that is, 53 (participants) * 2 (body sides) * 2 (maximum possible pROM score) = 212.

Dystonia and choreoathetosis scores were transformed into percentage scores relevant to their maximum score (maximum possible score of 288 for each subscale). A median percentage score and an interquartile range was calculated for the sample.

Statistical Analyses

The Shapiro-Wilk test showed that most data were not normally distributed, therefore non-parametric statistics were applied. Descriptive statistics were used to describe presence, severity, and distribution of spasticity, pROM deviations, dystonia, and choreoathetosis across body regions. The Wilcoxon-Signed Rank test was used for statistical comparisons of spasticity, pROM deviations, dystonia, and choreoathetosis (both between and within body regions).[33] Spearman's rank correlation coefficients (r_s) were used to explore the correlations between spasticity, pROM deviations, dystonia, choreoathetosis, and the functional classification scales (i.e. LL values for all parameters were correlated with the GMFCS whereas UL values for all parameters were correlated with the MACS). Correlation coefficients of <0.25 were considered as weak or no association, 0.25–0.50 as fair, 0.50–0.75 as moderate to good and >0.75 as excellent.[5] Statistical significance was set at $p<0.05$. Data was analysed using IBM SPSS Statistics v26 (SPSS Inc., Chicago, IL).

Results

Participants

Fifty-three participants (29 male), aged 6 to 22 years (mean age 15y 7mo, SD 4y 4mo) were included in this study. Ten participants were classified as GMFCS level I, five as level II, five as level III, seven as level IV and 26 as level V. Eight participants were classified as MACS level I, five as level II, six as level III, ten as level IV and 24 as level V. Signs of coexisting spasticity, pROM-, and pROM+ in at least one body region were present in 50 (94,3% of the sample), 52 (98,1% of the sample) and 53 participants (100% of the sample), respectively.

MAS presence and severity, pROM deviations, dystonia, and choreoathetosis

Median percentage values, statistical comparisons, and correlations of the MAS presence and severity, pROM deviations, dystonia, and choreoathetosis between body regions are presented in Table 1. The distribution across body regions is presented in Table 2 (absolute values and percentage values of MAS presence; absolute values, percentage values, and mode of MAS severity) and Table 3 (pROM- and pROM+).

Median percentage scores for the UL and the LL

The median percentage score of MAS presence for the UL was 16.7% (IQR 8.3–41.7%) and ranged between 0.0% and 75.0% of the maximum score, and for the LL was 50.0% (IQR 30.0-80.0%) ranging between 0.0% and 100% of the maximum score. The median percentage score of MAS severity for the UL was 6.3% (IQR 2.1-13.5%; range 0.0% and 30,2% of the maximum score) and for the LL was 18.8% (IQR 8.8-30.0%; range between 0.0% and 95.0%).

For pROM- deviations, the median percentage score for the UL was 8.3% (IQR 2.8-16.7%; range between 0.0% and 38.9% of the maximum score) and for the LL was 27.5% (IQR 15.0-35.0%; range between 0.0% and 62.5% of the maximum score). For pROM+ deviations, the median percentage score for the UL was 30.6% (IQR 22.2-36.1%; range between 2.8% and 55.6% of the maximum score) and for the LL was 17.5% (IQR 10.0-22.5%; range between 0.0% and 42.5%).

The median percentage score of dystonia for the UL was 83.3% (IQR 62.5–94.8%; range between 0.0%.9% and 100.% of the maximum score) and for the LL was 78.1% (IQR 52.1-87.5%; range between 21.9% and 100% of the maximum score). The median percentage score of choreoathetosis for the UL was 28.1% (IQR 14.6-53.1%; range between 0.0% and 89.6% of the maximum score) and for the LL was 18.8% (IQR 8.8-41.3%; range between 0.0% and 68.8% of the maximum score).

Distribution of MAS presence and severity, and pROM deviations across body regions

A high MAS presence was found in the plantar flexion muscles, that is, from the total 106 separate muscles (i.e. 53 participants * 2 body parts), 67 (63.2%) of total m.soleus and 75 (70.8%) of total m.gastrocnemius showed signs of spasticity presence. The lowest MAS presence was obtained in the forearm pronators with 6/106 (5.7%) and wrist palmarflexors with 5/106 (4.7%). MAS severity was higher in the plantar flexion muscles with a score of 110 (25.9%) and 136.5 (32.2%) out of the maximum severity score (i.e. 424) for m.soleus and m.gastrocnemius respectively, and lower in the palmar flexion muscles with a score of 4/424 (1.7%).

pROM- was more present and severe for knee extension (48.1% of the maximum score) in the LL, and shoulder internal rotation (35.8% of the maximum score) for the UL. On the other hand, pROM+ was more present and severe for hip internal rotation for the LL and shoulder external rotation for the UL with 44.8% and 47.2% of the maximum score respectively.

Statistical comparisons and correlations between and within body regions

Statistical comparisons revealed significantly higher levels of MAS presence, MAS severity, and pROM- in the LL compared to the UL with $p<0.001$. On the other hand, pROM+ ($p<0.001$), dystonia ($p=0.014$), and choreoathetosis ($p<0.001$) showed significantly higher levels in the UL compared to the LL. MAS presence and MAS severity were significantly

higher in the UL proximal (p=0.001 and p=0.002 respectively), and in the LL distal (p<0.001) compared to their counterparts. pROM- was significantly higher in the UL proximal (p<0.001) and LL proximal (p=0.004), whereas pROM+ was significantly higher in the UL distal (p<0.001) and in the LL distal with a difference not statistically significant (p=0.082). Dystonia was significantly higher in UL distal (p<0.001) and in the LL distal (p<0.001) whereas choreoathetosis levels showed no significant difference between proximal and distal parts of the UL nor LL.

The UL and the LL were significantly correlated for MAS presence, MAS severity, pROM-, pROM+, dystonia and choreoathetosis, with correlation coefficients ranging between $0.34<r_s<0.68$, ($p<0.001$ to $p=0.013$). Statistically significant correlations were also found between the UL proximal and UL distal for MAS presence, MAS severity, pROM-, dystonia, and choreoathetosis with correlation coefficients ranging between $0.32<r_s<0.76$ ($0.018<p<0.001$), as well as between LL proximal and LL distal for MAS presence, MAS severity, pROM-, pROM+, dystonia, and choreoathetosis ($0.32<r_s<0.63$, $0.020<p<0.001$).

MAS presence and severity, and pROM deviations related to dystonia and choreoathetosis

Correlation coefficients with 95% CIs and p-values of dystonia and choreoathetosis with presence of spasticity, severity of spasticity, and pROM deviations are presented in Table 4.

Total UL DIS-D showed fair and statistically significant correlations with the total UL MAS presence ($r_s=0.41$, 95% CI 0.16–0.61; p=0.002), the total UL MAS severity ($r_s=0.44$, 95% CI 0.19–0.63; p=0.001), and the total UL pROM- ($r_s=0.47$, 95% CI 0.23–0.66; p<0.001). Total LL DIS-D showed fair and statistically significant correlations with the total LL MAS presence ($r_s=0.33$, 95% CI 0.07–0.55; p=0.016), with the total LL MAS severity ($r_s=0.41$, 95% CI 0.16–0.61; p=0.002), and the total LL pROM- ($r_s=0.31$, 95% CI 0.04–0.54; p=0.025).

Total UL DIS-CA showed no statistically significant correlations with the total UL MAS presence, total UL MAS severity or total UL pROM deviations. Total LL DIS-CA showed fair

and statistically significant correlations only with the total LL pROM+ (r_s=0.44, 95% CI 0.19–0.63; p=0.001).

MAS presence and severity, pROM deviations, dystonia and choreothetosis related to the GMFCS and the MACS

Correlation coefficients with 95% CIs and p-values of MAS presence and severity, pROM deviations, dystonia and choreoathetosis with the GMFCS and the MACS are presented in Table 5.

The GMFCS showed a fair and statistically significant correlation with both the MAS presence score (r_s=0.32; 95% CI 0.06–0.54; p=0.018) and the MAS severity score (r_s=0.42; 95% CI 0.17–0.62; p=0.002) whereas moderate to good and statistically significant correlation was obtained with the pROM- (r_s=0.51; 95% CI 0.28-0.69; p<0.001) and DIS-D (r_s=0.65; 95% CI 0.46-0.78; p<0.001).

The MACS showed fair and statistically significant correlations with the MAS presence score (r_s=0.40; 95% CI 0.15–0.60; p=0.003), the MAS severity score (r_s=0.44; 95% CI 0.19–0.63; p=0.001), the pROM- (r_s=0.34; 95% CI 0.08–0.56; p=0.014), and DIS-CA (r_s=0.31; 95% CI 0.04–0.54; p=0.027). The MACS showed excellent and statistically significant correlation with the DIS-D (r_s=0.78; 95% CI 0.65–0.87; p<0.001).

Discussion

The overall aim of this study was to increase insights in the presence, severity, and distribution of spasticity and pROM deviations in individuals with DCP, and to assess their relationship with dystonia, choreoathetosis, and functional classification scales.

The current study is the first to differentiate between the presence and severity of the coexisting signs of spasticity in children with DCP. Obtained median percentage values for the UL and the LL show a higher presence of spasticity compared to its severity, which spasticity severity

is clearly lower than the severity of dystonia and choreoathetosis (except for the distal LL). Overall, these results are not unexpected given that a DCP diagnosis is assigned when dystonia and choreoathetosis are the dominant movement disorders,[1] however, notable for not being previously reported. 94.3% of the included participants showed signs of spasticity in at least one muscle group. A population-based study and a more recent cross-sectional study stated that 69% and 71% of individuals with DCP show signs of coexisting spasticity, respectively.[11,12] These studies do not report the muscle groups that were included in measuring coexisting spasticity in DCP, thus making it difficult to compare results with the current study. Higher presence and severity of spasticity were obtained for the LL whereas dystonia and choreoathetosis were more present and severe in the UL, which aligns with previous research.[11,34] Interestingly, spasticity and limited pROM showed fair and significant correlations with dystonia for the proximal LL but not for the distal LL (where the highest spasticity severity occurs). Correlation coefficients are greater if there is more variability among the observations.[35] The interquartile range in the current study shows a lower variability among the observations in the distal LL compared to the proximal LL, especially noticeable for dystonia. The lower variability in dystonia values may thereby explain the lack of significant correlations with spasticity and limited pROM of the distal LL. Nevertheless, the current study is an added value to the body of literature as it extensively describes the distribution of coexisting spasticity across body regions in DCP. This is of importance because managing strategies in CP differ among the various movement disorders and their characteristics.[8] For instance, botulinum toxin type A (BoNT-A) and intrathecal baclofen show promising results in reducing spasticity, dystonia, and in increasing ROM in DCP.[2,36-38] On the other hand, specific treatments like selective dorsal rhizotomy would reduce spasticity but have no impact on reducing dystonia[8,14,37], whereas deep brain stimulation is most efficient in individuals who have prominent dystonia and none to minimal presence of spasticity.[39]

The current study found fair and significant correlations for spasticity and moderate to good/excellent and significant correlations for dystonia when correlated to the GMFCS and the MACS levels. This may indicate that spasticity has a negative impact on the functional abilities, but perhaps to a lesser extent than dystonia.[6] Previous research suggests that the severity of dystonia may be such that any features of coexisting spasticity may be overlooked.[8] When treatment focuses on reducing severe dystonia, an adverse effect could include worsening of choreoathetosis, which indicates that dystonia may prevent the full expression of choreoathetosis.[2] Thus similarly, it may be that if treatment focuses on reducing severe dystonia while coexisting spasticity is left untreated, spasticity may surface and have a larger negative impact on function. The obtained insights in the current study on the presence, severity, and the distribution of dystonia, choreoathetosis, spasticity, and pROM deviations in DCP are clinically important to inform treatment management and ensure better clinical outcomes. Future research exploring more in-depth the clinical implications of this coexistence are recommended.

This is the first study to map pROM deviations of upper and lower limbs in participants with DCP during 38 joint movements, and particularly novel in differentiating between limited- and increased pROM. The hypertonic components of dystonia and spasticity combined with limited pROM lead to fixed musculoskeletal deformities (FMDs)[16] which are often associated with pain and impaired functioning in children and adolescents with DCP.[40] In the current study, this is supported by the fair significant correlations of dystonia with spasticity and limited pROM but not with hypermobility, likely explained by the hypertonic characteristics of both dystonia and spasticity.[3] A previous study reported a high occurrence of FMDs in DCP (accounting for 58% of the included CP cohort), particularly characteristic in DCP cases with mixed hypertonia.[16] FMD onset in DCP cases where spasticity coexists occurs typically around the age of four, which is significantly earlier than in pure DCP cases with an FMD onset at age

316 of nine.[16] The FMDs were not explored in the current study, however, the obtained high
317 percentage of coexisting spasticity and pROM deviations strengthens the claim that individuals
318 with DCP are highly prone to develop FMDs. Given their link with pain and impaired
319 functioning, these additional insights on the distribution of limited pROM across body regions
320 might assist clinical practice with a preventive treatment to maintain normal ROM and
321 symmetric postures at the earliest age possible. For instance, the current study found a higher
322 percentage of limited pROM for hip adduction, hip external rotation, and hip flexion. Previous
323 studies also report a higher presence of musculoskeletal deformities in the hip region compared
324 to other body regions in patients with DCP.[16] Moreover, a population-based study also reported
325 that children with spastic CP show lower ROM for hip adduction, hip external rotation, and hip
326 flexion when compared to typically-developing children.[17] Thus, it may be plausible that the
327 hip limited pROM obtained in the current study is due to the coexistence of spasticity which
328 causes limited pROM in the hip region in people with DCP. Structural changes happen in the
329 hips due to imbalances between flexor/extensor and adductor/abductor muscles, and because
330 of decreased muscle extension and decreased joint ROM.[41] Painful hip dislocation is a common
331 occurrence in CP, the incidence of which can be up to 80% of CP cases and is directly related
332 to the severity of the neurological involvement as well as the ambulatory status of the
333 patient.[40,42-45] Previous study reports an overall intermediate risk for hip dislocations in people
334 with DCP, however, up to 20% were classified in the highest risk group and this incidence was
335 directly related to their GMFCS levels.[46] Another study reported that the prognosis for hip
336 displacement was the worse in DCP with 24% of the included sample having abnormal hips.[47]
337 In addition, a high percentage of limited pROM in DCP was also found for shoulder internal
338 rotation, which aligns with previous research reporting structural changes at the shoulder level,
339 especially in mixed cases of CP.[48] The obtained distribution of the limited pROM in DCP is of
340 particular clinical interest given that significant associations were found between limited

pROM, the GMFCS, and the MACS. This might mean that treatment should specifically focus on increasing pROM values at the hip and shoulder level in order to improve functional abilities and/or reduce pain in DCP.

In the current study, joint hypermobility (i.e. increased pROM) in at least one joint movement was obtained for all participants, with a higher percentage for hip internal rotation, shoulder adduction, and shoulder external rotation. Increased pROM in DCP has not been described before, thus results cannot be compared to existing literature. Nonetheless, the fair significant correlations which were obtained between joint hypermobility and choreoathetosis are worth noting. Previous research reports joint hypermobility alongside hypotonia,[49] and maybe similarly, the underlying hypotonic and hyperkinetic (i.e. stormy and high amplitude movements) characteristics of choreoathetosis align with increased pROM in DCP.[4] Clinical practice supports there is a high luxation risk for individuals with DCP. Even though the current study found no significant correlations between joint hypermobility and functional motor abilities (i.e. the GMFCS and the MACS), future research is strongly encouraged to explore more in-depth its clinical implications, especially because in other populations, joint hypermobility has been associated with chronic joint pain, joint subluxations and dislocations, and soft tissue injuries.[49]

Although with clinically important findings to be reported, this study warrants some critical reflections. First, the assessments used in the current study are widely used in clinical research, and their reliability and validity has been reported before.[50-52] The insights generated from these studies have increased our understanding of spasticity and ROM, but the interpretation of the scales is dependent on the rater's execution, which could lead to subjective conclusions. In response to this, these evaluation methods are often complemented by the use of sensor measurements and wearables like electromyography recordings, 3D motion analysis or inertial measurement units.[53] Future research including sensor measurements and wearables may lead

to more in-depth insights on the clinical presentation of all movement disorders and impairments in DCP. In addition to their use in clinical research, there is also a need to bridge the implementation of these sensor measurements in clinical practice. Second, based on the International Classification of Functioning, Disability and Health (ICF) model[54], clinical outcomes should be assessed on levels of body functioning and structure, activity, and participation. The present study reported the associations between coexisting spasticity and pROM deviations with functional abilities such as the gross motor and fine manual abilities (i.e. body functioning and structure level), thus future research is necessary to explore their implications on other domains such as activity and participation. Third, as indicated by the fair positive correlations between coexisting spasticity and limited pROM with functional scales, it might be possible that there is a slight overestimation of their presence and severity in the UL and LL for the whole population due to the higher number of participants with GMFCS and MACS levels V. However, it is expected this effect to be very minimal given the only fair correlations and wide variability within the functional levels. Lastly, discussing the obtained results by comparing them with previous literature is necessary as it places the study in the field of research. However, it is worth highlighting that the differences which exist between the current study and previous relevant studies (differences in methodology including sample size, assessed body regions, recruitment procedures and inclusion criteria among others) make the comparison of the obtained results more difficult and should therefore be considered.

Conclusion

This study aimed to increase insights in the clinical presentation of spasticity and pROM deviations alongside dystonia and choreoathetosis in individuals with DCP and to assess their relationship with dystonia, choreoathetosis and functional abilities. The severity of spasticity is lower than the severity of dystonia and choreoathetosis, thus dystonia and choreoathetosis remain the dominant movement disorders in DCP. Given the obtained correlations with the

functional scales, spasticity and limited pROM seem to have functional implications and should be therefore addressed accordingly. Hypermobility, on the other hand, may lead to an increased luxation risk. The obtained insights in the current study on the presence, severity, and the distribution of dystonia, choreoathetosis, spasticity, and pROM deviations in DCP should inform treatment management and ensure better clinical outcomes.

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Declaration of interest

The authors report no conflicts of interest.

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Table 1: Median and interquartile range of the presence and severity of spasticity, passive range of motion deviations, dystonia, and choreoathetosis; statistical comparisons and correlation coefficients between and within body regions

	Median (IQR) (%)	Median (IQR) (%)	Wilcoxon	r_s (95% CI), p-value
	Total UL	**Total LL**		
MAS presence	16.7 (8.3–41.7)	50.0 (30.0–80.0)	<0.001	0.34 (0.08–0.56), p=0.013
MAS severity	6.3 (2.1–13.5)	18.8 (8.8–30.0)	<0.001	0.38 (0.12–0.59), p=0.005
pROM-	8.3 (2.8–16.7)	27.5 (15.0–35.0)	<0.001	0.59 (0.38–0.74), p<0.001
pROM+	30.6 (22.2–36.1)	17.5 (10.0–22.5)	<0.001	0.44 (0.19–0.63), p=0.001
DIS-D	83.3 (62.5–94.8)	78.1 (52.1–87.5)	0.014	0.68 (0.50–0.80), p<0.001
DIS-CA	28.1 (14.6–53.1)	18.8 (8.8–41.3)	<0.001	0.68 (0.50–0.80), p<0.001
	UL proximal	**UL distal**		
MAS presence	25.0 (0.0–50.0)	12.5 (0.0–25.0)	0.001	0.47 (0.23–0.66), p<0.001
MAS severity	9.4 (0.0–25.0)	4.7 (0.0–9.4)	0.002	0.53 (0.30–0.70), p<0.001
pROM-	10.7 (3.6–17.9)	0.0 (0.0–0.0)	<0.001	0.32 (0.06–0.54), p=0.018
pROM+	21.4 (17.9–32.1)	50.0 (37.5–50.0)	<0.001	0.22 (-0.05–0.46), p=0.100
DIS-D	83.3 (58.3–95.8)	93.8 (66.7–97.9)	<0.001	0.64 (0.45–0.78), p<0.001
DIS-CA	31.3 (10.4–58.3)	28.1 (14.6–50.0)	0.870	0.76 (0.62–0.85), p<0.001
	LL proximal	**LL distal**		
MAS presence	33.3 (0.0–66.7)	75.0 (50.0–100.0)	<0.001	0.47 (0.23–0.66), p<0.001
MAS severity	10.4 (0.0–25.0)	25.0 (12.5–37.5)	<0.001	0.50 (0.27–0.68), p<0.001
pROM-	28.1 (18.8–40.6)	25.0 (0.0–25.0)	0.004	0.51 (0.28–0.69), p<0.001
pROM+	15.6 (9.4–21.9)	25.0 (0.0–37.5)	0.082	0.32 (0.06–0.54), p=0.020
DIS-D	68.8 (31.3–81.3)	83.3 (72.9–93.8)	<0.001	0.52 (0.29–0.69), p<0.001
DIS-CA	21.9 (4.2–37.5)	20.8 (4.2–43.8)	0.213	0.63 (0.43–0.77), p<0.001

MAS, Modified Ashworth Scale; pROM-, reduced passive range of motion; pROM+, increased passive range of motion (hypermobility); DIS-D, Dyskinesia Impairment Scale – Dystonia subscale; DIS-CA, Dyskinesia Impairment Scale – Choreoathetosis subscale; UL, upper limbs; LL, lower limbs; IQR, Interquartile Range; Wilcoxon, Wilcoxon signed-rank test; r_s, Spearman's rho correlation coefficient; CI, confidence interval

Table 2: Absolute values (summed score out of the maximum score) and percentage values of the presence and severity of spasticity across different joint movements and muscle groups

			MAS presence (summed score/maximum score)	MAS presence (%)	MAS severity (summed score/maximum score)	MAS severity (%)	MAS severity (mode of MAS score, excluding 0 scores)
LL	Ankle	Plantar flexion (soleus)	67/106	63.2	110/424	25.9	1.5
		Plantar flexion (gastrocnemius)	75/106	70.8	136.5/424	32.2	2
	Hamstrings	Popliteal angle	55/106	51.9	83.5/424	19.7	1
	Hip	Abduction (90°)	36/106	34.0	62/424	14.6	1.5
		Abduction (0°)	37/106	34.9	58.5/424	13.8	1.5
UL	Elbow	Flexion (145°)	40/106	37.7	69.5/424	16.4	1
		Extension (0°)	30/106	28.3	50/424	12.5	1
	Forearm	Supination (85°)	33/106	31.1	48.5/424	11.4	1
		Pronation (90°)	6/106	5.7	9.5/424	2.2	1
	Wrist	Dorsiflexion (70°)	24/106	22.6	36.5/424	8.6	2
		Palmar flexion (80°)	5/106	4.7	7/424	1.7	1

LL, lower limbs; UL, upper limbs; MAS, Modified Ashworth Scale; %, percentage;

Table 3: Percentage values of passive range of motion deviations across different joint movements; median and interquartile range of degrees of deviation

			Normal pROM	pROM –		pROM +	
			(degrees)	%	Median (IQR) (degrees)	%	Median (IQR) (degrees)
LL	Ankle	Dorsiflexion	20°	10.8	-10 (-10–-10)	28.8	15 (10–25)
		Plantarflexion	45°	32.1	-10 (-10–-5)	13.2	10 (8.8–15.0)
	Knee	Flexion	145°	14.2	-15 (-35–-10)	21.2	15 (5–15)
		Extension	5°	48.1	-15 (-25–-5)	2.8	5 (5–5)
	Hip	Abduction	45°	28.3	-10 (-15–-5)	23.6	10 (5–15)
		Adduction	30°	34.0	-10 (-15–-10)	2.4	10 (5–10)
		Flexion	125°	39.6	-15 (-25–-5)	12.3	5 (5–10)
		Extension	10°	19.8	-10 (-20–-5)	15.1	10 (5–10)
		External rotation	45°	33.5	-15 (-25–-10)	15.6	5 (5–15)
		Internal rotation	30°	2.8	-10 (-13.8–-6.3)	44.8	30 (20–40)
UL	Elbow	Flexion	145°	13.7	-5 (-5–-5)	36.3	5 (5–10)
		Extension	0°	5.7	-10 (-10–-10)	34.4	10 (5–10)
	Shoulder	Flexion	180°	17.9	-30 (-45–-20)	0.0	–
		Abduction	180°	17.5	-30 (-40–-20)	0.9	20 (20–20)
		Adduction	30°	2.4	-5 (-6.3–-5)	41.0	15 (10–20)
		External rotation	80°	1.4	-30 (-35–-20)	47.2	10 (10–10)
		Internal rotation	70°	35.8	-20 (-30–-10)	6.6	10 (8.8–20)
	Wrist	Dorsiflexion	70°	6.6	-15 (-42.5–-10)	41.0	20 (10–20)
		Palmar flexion	80°	3.3	-10 (-12.5–-10)	50.9	20 (10–25)

LL, lower limbs; UL, upper limbs; pROM-, limited passive range of motion; pROM+, increased passive range of motion (hypermobility); %, percentage, IQR, interquartile range

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Table 4: Correlation coefficients of dystonia and choreoathetosis with presence of spasticity, severity of spasticity, and passive range of motion deviations

			MAS presence r_s (95% CI); p-value	MAS severity r_s (95% CI); p-value	pROM- r_s (95% CI); p-value	pROM+ r_s (95% CI); p-value
DIS-D						
	UL	Total	0.41 (0.16–0.61); p=0.002	0.44 (0.19–0.63); p=0.001	0.47 (0.23–0.66); p<0.001	-0.09 (-0.35–0.18); p=0.538
		Proximal	0.50 (0.27–0.68); p<0.001	0.54 (0.32–0.71); p<0.001	0.37 (0.11–0.58); p=0.007	-0.06 (-0.33–0.21); p=0.674
		Distal	0.17 (-0.11–0.42); p=0.213	0.18 (-0.09–0.43); p=0.210	0.35 (0.09–0.57); p=0.011	0.03 (-0.24–0.30); p=0.853
	LL	Total	0.33 (0.07–0.55); p=0.016	0.41 (0.16–0.61); p=0.002	0.31 (0.04–0.54); p=0.025	0.05 (-0.22–0.32); p=0.730
		Proximal	0.37 (0.11–0.58); p=0.007	0.45 (0.21–0.64); p=0.001	0.35 (0.09–0.57); p=0.010	-0.05 (-0.17–0.37); p=0.762
		Distal	0.18 (-0.09–0.43); p=0.191	0.18 (-0.09–0.43); p=0.207	0.15 (-0.13–0.40); p=0.285	0.11 (-0.05–0.46); p=0.439
DIS-CA						
	UL	Total	0.26 (-0.01–0.50); p=0.064	0.22 (-0.05–0.46); p=0.121	0.14 (-0.14–0.40); p=0.320	0.07 (-0.20–0.30); p=0.631
		Proximal	0.35 (0.09–0.57); p=0.010	0.35 (0.09–0.57); p=0.010	0.08 (-0.19–0.34); p=0.589	0.11 (-0.17–0.37); p=0.449
		Distal	-0.03 (-0.30–0.24); p=0.839	-0.07 (-0.33–0.20); p=0.605	0.05 (-0.22–0.32); p=0.746	0.04 (-0.23–0.31); p=0.768
	LL	Total	0.15 (-0.13–0.40); p=0.294	0.14 (-0.14–0.40); p=0.320	-0.25 (-0.49–0.02); p=0.077	0.44 (0.19–0.63); p=0.001
		Proximal	0.26 (-0.01–0.50); p=0.062	0.26 (-0.01–0.50); p=0.063	-0.04 (-0.31–0.23); p=0.806	0.35 (0.09–0.57); p=0.012
		Distal	0.05 (-0.22–0.32); p=0.719	0.01 (-0.26–0.28); p=0.960	-0.26 (-0.50–-0.01); p=0.058	0.18 (-0.09–0.43); p=0.202

DIS-D, Dyskinesia Impairment Scale – Dystonia subscale; DIS-CA, Dyskinesia Impairment Scale – Choreoathetosis subscale; UL, upper limbs; LL, lower limbs; MAS, Modified Ashworth Scale; pROM-, limited passive range of motion; pROM+, increased passive range of motion (hypermobility); r_s, Spearman's rho correlation coefficient; CI, confidence interval

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Table 5: Correlation coefficients of the Gross Motor Functional Classification System with the presence of spasticity, severity of spasticity, passive range of motion deviations, dystonia, and choreoathetosis of the lower limbs; and correlation coefficients between the Manual Ability Classification System with the presence of spasticity, severity of spasticity, passive range of motion deviations, dystonia, and choreoathetosis of the upper limbs

	GMFCS		**MACS**	
	r_s (95% CI)	***p* value**	**r_s (95% CI)**	***p* value**
MAS presence	0.32 (0.06–0.54)	0.018	0.40 (0.15–0.60)	0.003
MAS severity	0.42 (0.17–0.62)	0.002	0.44 (0.19–0.63)	0.001
pROM-	0.51 (0.28–0.69)	<0.001	0.34 (0.08–0.56)	0.014
pROM+	-0.23 (-0.47–0.04)	0.099	-0.10 (-0.36–0.18)	0.490
DIS-D	0.65 (0.46–0.78)	<0.001	0.78 (0.65–0.87)	<0.001
DIS-CA	0.07 (-0.20–0.33)	0.632	0.31 (0.04–0.54)	0.027

GMFCS, Gross Motor Function Classification System; MACS, Manual Ability Classification System; MAS, Modified Ashworth Scale; pROM-, reduced passive range of motion; pROM+, increased passive range of motion (hypermobility); DIS-D, Dyskinesia Impairment Scale – Dystonia subscale; DIS-CA, Dyskinesia Impairment Scale – Choreoathetosis subscale; r_s, Spearman's rho correlation coefficient; CI, confidence interval

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