Lower-extremity selective voluntary motor control in patients with spastic cerebral palsy: increased distal motor impairment

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LIST OF ABBREVIATIONS

CST	Corticospinal tract
PDC	Proximal to distal concordance
SCALE	Selective Control Assessment of the Lower Extremity
SVMC	Selective voluntary motor control

AIM Multiple impairments contribute to motor deficits in spastic cerebral palsy (CP). Selective voluntary motor control (SVMC), namely isolation of joint movement upon request, is important, but frequently overlooked. This study evaluated the proximal to distal distribution of SVMC impairment among lower extremity joints.
METHOD Using a recently developed tool, the Selective Control Assessment of the Lower Extremity (SCALE), we evaluated the SVMC of the hip, knee, ankle, subtalar joint, and toes in a cross-sectional, observational study of 47 participants with spastic, diplegic, hemiplegic, and quadriplegic CP (22 males, 25 females; mean age 11y 9mo, SD 4y 8mo; Gross Motor Function Classification System levels I–IV).
RESULTS Statistically significant decreases in SCALE scores from hip to toes were found using the Page statistical test for trend ($p<0.001$). Statistically significant differences ($p<0.05$) were found between all joint pairs, except toes versus subtalar, toes versus ankle, and right ankle versus subtalar joints. Cross-tabulation of score frequencies for all pairs revealed that proximal joint scores were higher or equal to distal ones 81 to 100% of the time. Excluding toes versus subtalar joints, proximal scores exceeded distal ones 94 to 100% of the time.
INTERPRETATION We confirmed increasing proximal to distal SVMC impairment, which may have implications for treatment and research.

Individuals with cerebral palsy (CP) have limitations in motor function resulting from multiple impairments including spasticity, contractures, weakness, and diminished selective motor control. Selective motor control has been defined as '… the ability to isolate the activation of muscles in a selected pattern in response to demands of a voluntary movement or posture.'[1] Selective voluntary motor control (SVMC) describes the performance of specific isolated joint movements upon request, as opposed to the habitual activation of selected muscles during functional tasks.[2] SVMC at the ankle is a strong predictor of functional movement ability in children with CP,[3] and SVMC has been used as a prognostic factor in selecting candidates for selective posterior rhizotomy.[4] We recently reported the development, validity, and reliability of the Selective Control Assessment of the Lower Extremity (SCALE).[2] SCALE is a clinical tool designed to assess SVMC of the entire lower extremity by summing the scores for five joints (hip, knee, ankle, subtalar joint, and toes).

Voluntary isolated joint movements require activation of the corticospinal tracts (CSTs). In patients with CP, damage to these tracts commonly occurs in the periventricular area.[5] Damage to the periventricular white matter is the most frequent abnormal magnetic resonance imaging (MRI) finding associated with the spastic diplegic form of CP and it is found in more than one-third of those with hemiplegia and quadriplegia.[6] Strong correlations between damage to the CSTs and motor impairment have been reported for children with CP.[5,7]

The CSTs have a specific anatomical arrangement as they descend from the motor cortex to the spinal motor neuron pools.[8–10] In the periventricular area, motor fibers leading to the lower extremities are more likely to be damaged than those supplying the upper extremities because of their more medial position.[10] This anatomical relationship has been confirmed in recent studies using MRI tractography.[11,12] The somatotopic organization of the lower extremity in the sensorimotor cortex suggests that distal lower-extremity tracts are closer to the ventricle and more vulnerable than those of proximal lower-extremity muscles (Fig. 1). Evidence exists of increased distal impairment of lower-extremity motor function in children with spastic CP,[13–15] but studies specifically examining the relationship between SVMC of proximal compared with distal lower-extremity joints have not been reported. Tedroff et al.[13] evaluated the temporal sequence of muscle recruitment during maximal voluntary contractions in patients with

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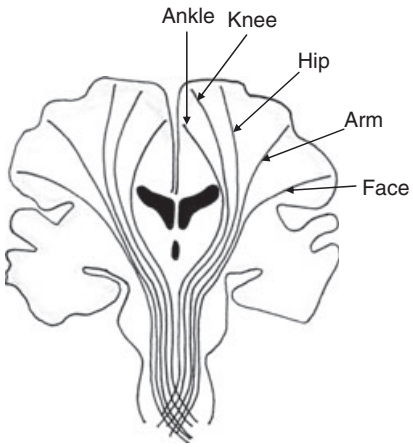

Figure 1: Representation of corticospinal tracts as they relate to lateral ventricles (adapted from Aicardi and Bax[10]).

Table I: Participant characteristics (*n*=47)

Age (y:mo)	
Mean (SD)	11:9 (4:8)
Range	5:1–23:0
Characteristic (*n*)	
Sex	
Male	22
Female	25
Distribution of impairment	
Diplegia (three had hemiplegic overlay)	35
Hemiplegia	6
Quadriplegia	6
GMFCS-ER level	
I	10
II	10
III	18
IV	9

GMFCS-ER, Gross Motor Function Classification Scale, Expanded and Revised edition.

hemiplegic and diplegic CP compared with participants without disability. Inappropriate activation of non-agonists before agonists was more prevalent in distal than proximal musculature in children with CP. During gait, Wakeling et al.[14] reported that disordered muscle firing occurred more frequently in distal than proximal musculature in children with spastic diplegia. In addition, greater muscle weakness has been quantified at the ankle joint than at more proximal joints.[15–17]

The aim of this study was to analyse the distribution of SVMC scores among lower-extremity joints in patients with spastic CP using the SCALE tool. We hypothesized that SVMC impairment would be greater in distal than proximal joints.

METHOD

This cross-sectional, observational study was approved by the University of California, Los Angeles, institutional review board. All participants with CP, or their parent or legal guardian, provided informed assent and consent. Forty-seven individuals with spastic CP who attended the UCLA/Orthopaedic Hospital Center for Cerebral Palsy interdisciplinary clinic in Los Angeles volunteered to participate. Consecutive individuals who met inclusion criteria were invited to enroll. It was important to include participants across the spectrum of severity, based on Gross Motor Function Classification System (GMFCS) level. There were a minimum of nine participants representing each GMFCS level (I–IV). Previous work revealed that the SCALE assessment could not be performed for patients at GMFCS level V.[2] This sample size was considered sufficient, based on previous work showing significant correlation between GMFCS levels and SCALE scores for 51 participants.[2] Participant characteristics are presented in Table I. The following inclusion criteria were used: (1) diagnosis of spastic CP, (2) ability to follow simple directions, and (3) age between 4 and 25 years. The following exclusion criteria were used: (1) history of lower-extremity musculotendinous transfer or joint fusion, (2) neurosurgical or musculoskeletal surgery within the past year, (3) initial placement of baclofen pump within the past year, (4) botulinum toxin injections within 5 months, or (5) musculoskeletal injury within the past month.

One of three experienced raters performed the SCALE assessment for the right and left lower extremity of each participant. These raters previously demonstrated a high level of interrater reliability with intraclass correlation coefficients ranging from 0.88 to 0.91.[2] Each participant was asked to perform specific non-synergistic reciprocal movements and scores of unable 0, impaired 1, or normal 2 were assigned for the hip, knee, ankle, subtalar, and toe joints for each side.[2] All tests were performed in the sitting position, with the exception of the hip test, which was performed side-lying with the limb supported by the examiner. The patient was asked to perform the following reciprocal movement patterns: (1) hip flexion and extension with the knee extended, (2) knee extension and flexion, (3) ankle dorsiflexion and plantarflexion with the knee extended, (4) subtalar inversion and eversion, and (5) toe flexion and extension. A normal score (2) was given when the participant demonstrated isolated reciprocal joint motion through at least 50% of the available passive range of motion within an approximately 3-second verbal count. Unable (0) was assigned if the participant could not move the joint or if the attempted movement occurred in a synergistic pattern (simultaneous one-to-one movement at two or more joints of the same limb). A grade of impaired (1) was given if one or more of the following occurred: (1) the range of active movement was less than 50% of the participant's available passive range of motion, (2) movement occurred in only one direction, (3) the task was performed slower than a 3-second verbal count, (4) motion at untested joints occurred (including mirror movement of the opposite limb). (See Data S1, the SCALE Score Sheet, Directions for Administration, and Instructions for Grading, supporting information, published online.)

Individual joint SCALE scores were compared using nonparametric repeated measures methods (Friedman procedure); the corresponding test for trend (Page test)[18] was computed to analyse the relation among joints from hip to toes for the left

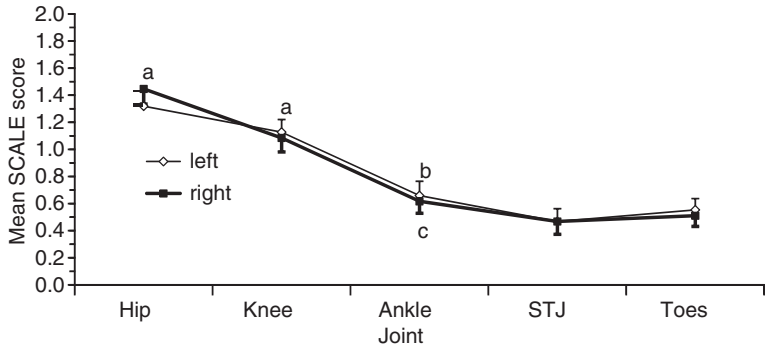

Figure 2: Mean Selective Control Assessment of the Lower Extremity (SCALE) scores by joint for left and right lower extremities. A score of 0.0 indicates the participant was unable to isolate or used the full synergy pattern, 1.0 indicates impaired motor control, and 2.0 indicates normal isolated movement. Error bars represent one standard deviation (+ for left and – for right). a, significant differences for all joint pairs on both left and right ($p<0.05$); b, left ankle score was significantly different from left hip, knee, and subtalar joint scores ($p<0.05$); c, right ankle joint score was significantly different from right hip and knee ($p<0.05$) and showed a tendency toward a difference from the subtalar joint score ($p=0.065$). Specific p values for all joint pairs are given in Table II. STJ, subtalar joint.

and right lower limbs. Additionally, score frequencies for all pairs of joints were cross-tabulated for the left and right sides. When the proximal joint score was equal to or greater than the distal joint, we called this proximal to distal concordance (PDC). The percentage of PDC was calculated for all combinations of joint pairs within each limb. One hundred per cent PDC indicated that distal joint scores never exceeded those of proximal joints. StatXact 8.0 (Cytel Inc, Cambridge, MA, USA) was used for statistical computations.

RESULTS

Mean SCALE scores showed greater SVMC impairment in distal than proximal joints bilaterally (Fig. 2). A statistically significant decrease in SCALE scores from hip to toes was found using the Page statistical test for trend ($p<0.001$). Significant differences were found between all pairwise SCALE score comparisons involving the hip and the knee joints bilaterally. The left ankle scores were significantly different from all other joint scores except the toes. The right ankle joint scores differed from the hip and knee scores and showed a tendency toward a difference from the subtalar joint score ($p=0.065$) Comparisons between scores for toes versus subtalar, and toes versus ankle, did not show a significant difference for either limb. Table II presents p values for all pairwise comparisons using the Friedman test. The percentage PDC for cross-tabulations of joint score frequencies ranged from 81 to 100% (Fig. 3). Excluding comparisons between score frequencies for toes and subtalar joints, the percentage PDC was 94 to 100% (Fig. 3). Distal joint scores exceeded proximal ones for only nine limbs (seven participants) when comparisons involving the toes were excluded.

DISCUSSION

To our knowledge, this is the first study to report SVMC of the hip, knee, ankle, subtalar, and toe joints in individuals with spastic CP. Our hypothesis of greater distal than proximal SVMC impairment within each limb was mostly confirmed.

Table II: Comparison between SCALE scores for five joints on the right and left with individual p values (Friedman test) and overall trend (Page test)

Joint comparison	Left p value	Right p value
Hip vs knee	0.023	<0.001
Hip vs ankle	<0.001	<0.001
Hip vs subtalar joint	<0.001	<0.001
Hip vs toes	<0.001	<0.001
Knee vs ankle	<0.001	<0.001
Knee vs subtalar joint	<0.001	<0.001
Knee vs toes	<0.001	<0.001
Ankle vs subtalar joint	0.023	0.065
Ankle vs toes	0.227	0.180
Subtalar joint vs toes	0.424	0.774
Overall trend	<0.001	<0.001

Significant at $p=0.023$ and $p<0.001$. SCALE, Selective Control Assessment of the Lower Extremity.

These results support the concept of increased vulnerability of CSTs associated with distal lower-extremity musculature. Previous research examining muscle strength[15–17] similarly found increased deficits in distal joints. Impaired SVMC may be associated with the observations reported in these studies. To determine the relative influence of muscle strength (force-generating capacity) and SVMC (neurological recruitment by CSTs) on movement production, both test positioning and the movement pattern requested must be examined. In designing the SCALE ankle assessment for isolated motion out of synergy, we positioned the knee in extension when requesting ankle dorsiflexion. To verify force-generating ability at the ankle, a flexor synergy pattern was elicited separately by resisting hip flexion and noting the active ankle dorsiflexion. This phenomenon has been referred to as the 'confusion test'.[19] Participants in the present study with an absence of SVMC at the ankle (a SCALE score of 0) could demonstrate active ankle dorsiflexion only when using the total flexor synergy pattern. Wiley and Damiano[15] found greater ankle dorsiflexor strength

Figure 3: Cross-tabulations of Selective Control Assessment of the Lower Extremity score frequencies for (a) left and (b) right lower-extremity joints showing the percentage proximal to distal concordance (PDC). Shaded zones indicate relationships that violated PDC because the distal joint scores exceeded the proximal joint scores. STJ, subtalar joint.

deficits when the knee was extended rather than flexed, demonstrating the influence of impaired SVMC. Although these investigators concluded that distal muscles were generally weaker than proximal muscles, the hip extensors were an exception, being weaker than the ankle muscles. As the SCALE hip test was performed in an antigravity side-lying position, minimizing the need for muscle force-generating capacity, we found that SVMC at the hip exceeded that found at the ankle.

Tedroff et al.[13] reported that during maximal voluntary contractions, children with spastic CP more frequently activated a muscle other than the intended prime mover first, especially when the prime mover was a more distal muscle. These results are consistent with our findings of decreased ability to perform isolated joint motion distally. As Tedroff et al. did not specify whether the participants performed isolated joint motion out of synergy, and we did not record electromyograms, direct comparisons are limited.

Excluding comparisons involving the toes, PDC exceptions in our study were rare, and could have been caused by scoring errors or other factors affecting patient performance, such

as impaired motor planning. Some exceptions to the PDC appeared to be related to the presence of either restricted range of motion or mirror movements. Contractures or severe spasticity can mask underlying SVMC. One 15-year-old participant with spastic diplegia (GMFCS level I) had a subtalar score of 2, whereas the more proximal ankle joint was scored 1, owing to restricted range of motion. As this study supports increased proximal SVMC, it is possible that this participant's ankle would have been scored 2 if tested at a younger age, before contracture development. The higher score at the subtalar joint predicts greater functional improvement after contracture release than if the score were 1 or 0. This is one example of how SVMC assessment, particularly as part of a periodic evaluation for children with spastic CP during development, may be helpful in predicting the potential for functional improvement after a specific treatment.

Mirror movements are simultaneous, obligatory movements that occur at contralateral joints during active movement. In adults without disability, most CSTs are crossed; however, extensive ipsilateral tracts are normally present in early development. In hemiparetic CP, there is evidence that surviving contralateral tracts may be competitively displaced by persistent ipsilateral tracts[20,21], which may be responsible for mirroring.[22] Using the SCALE tool, a score of 1 is given at a joint when the same movement pattern is observed contralaterally. Mirror movements negatively affected the PDC in the less involved limb for some participants with asymmetrical CP. In particular, two participants with spastic hemiplegia could isolate movement of their ankle joint on their non-hemiplegic side, but received scores of 1 owing to mirror ankle movement on their hemiplegic side. Mirroring did not occur during subtalar joint testing on their non-hemiplegic sides, giving these limbs a score of 2 at the more distal joint. Although the presence of mirroring is more likely to reflect a primary pathology for the hemiplegic limb, we assigned the SVMC impairment for the limb being assessed, as it is movement of this limb that elicits the abnormal movement pattern and any resulting functional problems. The effects of obligatory mirror movements on functional lower-extremity motor tasks such as walking are unknown and require further study.

The most frequent exceptions to the PDC occurred when the toes were graded as 1 and the subtalar joint was graded as 0, indicating absent subtalar SVMC with sparing at the toes. There are several possible explanations for these findings. One may be that the toes are not truly distal anatomically. Although the insertions of the toe musculature are more distal, the origin of muscles controlling the ankle, subtalar joint, and toes are similar. In addition, control of the subtalar joint appears to be more challenging than that of other joints. We observed that isolated motion of the subtalar joint was the most difficult movement sequence for participants with CP to understand and perform. Similar observations have been reported for patients after stroke. Eversion was described as a challenging movement in adults after stroke and is an indicator of the highest level of recovery for the lower extremity.[23] Another possible explanation is that moving only one of multiple toe joints was sufficient to obtain a SCALE score of 1, reducing the relative potential for a score of 0 at the toes compared with the subtalar joint. Finally, there may be greater capacity for sparing of corticospinal fibers associated with toe movement owing to greater density of CSTs. In early mapping studies of the human motor cortex, the area of cortical representation for the great toe was exceeded only by the tongue, mouth, thumb, and fingers.[8,9] More recent studies indicate that both toe musculature and tibialis anterior have a higher density of associated monosynaptic corticospinal projections than proximal lower-limb musculature.[24]

SVMC assessment and the proximal to distal distribution of impairment can be useful in treatment planning and in considering prognoses for the development of motor function in young children with CP. Based on the proximal to distal increase in SVMC impairment, patients who are assigned a score of 2 at the ankle or subtalar joint are more likely to have scores of 2 at the knee and hip. Although examining SVMC at the individual joint level can be helpful in treatment planning, the SCALE total limb score is more useful when describing a patient's overall functional ability. For example, we have shown that SCALE total limb scores are significantly related to the performance of simultaneous hip flexion and knee extension, as normally occurs, during the terminal swing phase of gait.[25]

We believe this is the first systematic evaluation and comparison of SVMC among multiple lower-extremity joints in individuals with spastic CP. It confirms the increase in severity of impairment from proximal to distal joints. Although previous research supports greater impairment in distal muscles and joints, this phenomenon has received little attention. Anatomical and physiological mechanisms contributing to these findings require further study. Our results support selective vulnerability of the corticospinal tracts innervating distal musculature owing to their proximity to the ventricles. Although the participants in this study had a clinical diagnosis of spastic CP, damage to the periventricular white matter was not documented in this sample. Newer technologies allow documentation of precise damage to white matter tracts using MRI with diffusion tensor imaging. This may be useful in elucidating the relation between the injury and functional impairment; these studies are currently in progress.

SUPPORTING INFORMATION

Additional supporting information is available for this article online:

Data S1: The Selective Control Assessment of the Lower Extremity (SCALE), Score Sheet, Directions for Administration and Instructions for Grading.

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