Original Article

Correlation of Range of Motion and Functional Performance of Upper Extremities in Children with Cerebral Palsy

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Objective: Upper extremity deformities in children with cerebral palsy greatly affects their functioning and their daily activities. Range of motion of the upper extremity joints in children with cerebral palsy is usually limited. There is little information available, however, regarding the relation between range of motion and functional outcomes of upper extremities in children with cerebral palsy. The purpose of this study was to determine the correlation both between the Upper Extremity Rating Scale [UERS] and the Manual Ability Classification System [MACS] and also between the UERS test and the Box and Block Test [BBT].

Materials and Methods: Fifty-seven children with cerebral palsy were enrolled in this cross-sectional study. Their range of motion was rated using the UERS, their ability to handle objects in daily activities was evaluated using MACS, and manual dexterity of the upper limbs was determined using BBT. The more-affected side of each child was evaluated. Correlations were assessed using Pearson's and Spearman's test.

Results: Pronation contracture and wrist flexion deformities were the most common problems in this study. Correlations in children with cerebral palsy between the UERS and BBT (Pearson's correlation, r = 0.5188; p<0.0001), and between UERS and MACS (Spearman's correlation, r = -0.5217; p<0.0001) were statistically significant.

Conclusion: There is a significant correlation between range of motion and functional performance of upper limbs in cerebral palsy patients. This suggests that strategies to improve in range of motion of upper limb joints should be emphasized in order to improve the functional performance in children with cerebral palsy.

Keywords: Cerebral palsy; Upper extremity deformity; Functional performance; Upper Extremity Rating Scale

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Cerebral palsy [CP] is a general term describing common features of disabilities and movement disorders resulting from static non-progressive brain damage during the developmental period in childhood. This condition may produce various neurological defects presenting as disturbances of sensation, perception, cognition, communication, behavior, and secondary musculoskeletal disorders(1). Defects of the lower extremities, including hip, knee and ankle, usually affect the ambulatory status of children with cerebral palsy. However, upper extremity deformities or contractures can have significant effects on patients' posture, motor function, and quality of life(2). The clinical presentation of upper extremity problems is heterogeneous, with variations in severity and pattern. Of cerebral palsy children, 83% have upper extremity problems(3,4). Common deformities include shoulder adduction and internal rotation, elbow flexion, forearm pronation, wrist

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and finger flexion, and thumb-in-palm deformity[2,5,6]. Forearm pronation and wrist flexion contractures are also common features, with a prevalence of 70.5% and 62.8%, respectively[4]. The magnitude of the problems not only affects the children's quality of life but also that of their parents and caregivers[7,8].

In cerebral palsy, the range of motion of shoulder, elbow and wrist are usually limited by spasticity, contracture, and/or joint deformity. The range of motion of joints is a product of both physical structure and performance. Individuals with chronic spasticity have been found to develop depletion of number of sarcomeres as well as having lower muscle growth and elasticity, which results in hypoextensibility of muscle[9-11]. As a consequence of spasticity and developmental disuse, cerebral palsy children may exhibit a decreased range of motion and stiffness of joints which affects upper extremity functions, including both the gross and fine hand motion required for manual ability and task completion[12]. Increasing range of motion is also one of the key goals in treating cerebral palsy, and physical therapy is a standard treatment for maintaining the motion of each joint[13,14].

It is believed that range of motion may be a key component in determining upper extremity function in children with cerebral palsy. The Manual Ability Classification System [MACS] and the Box and Block Test are widely used by both clinicians and healthcare providers for assessment of upper limbs in cerebral palsy cases. MACS is used for determining a child's typical manual performance for daily activities while the Box and Block Test [BBT] shows manual dexterity ability[15-17]. Until now, there has been little direct evidence of the association between the range of motion of the upper extremities and functional scores in children with cerebral palsy.

The purpose of this study was to determine the correlation between range of motion assessment by the Upper Extremity Rating Scale [UERS] and functional performance evaluation by the Manual Ability Classification System [MACS], as well as the correlation between UERS and BBT for the patients' more severely affected side. The significance of range of motion on the performance function in cerebral palsy patients is elucidated in the present study.

Materials and Methods

A total of 57 children with cerebral palsy who had attended in Srisangwan School for Disabled Children in Nonthaburi province, Thailand during the 2015 to 2016 school year were enrolled in the study. This cross-sectional study was conducted with the approval of the Committee on Human Rights Related to Research Involving Human Subjects, Faculty of Medicine, Ramathibodi Hospital, Mahidol University, Thailand (ID 05-59-01). Parents were informed about the study and gave their consent for participation. All subjects were willing to participate.

Participants were evaluated by orthopaedic surgeons and rehabilitation physicians for cerebral palsy type (anatomic and neuromuscular), dominant hand, Gross Motor Function Classification System [GMFCS] level (range: I-V), House functional classification system of hand function based on activity levels (range: 0- does not use to 8- spontaneous use, manipulating hand), Upper Extremity Rating Scale [UERS], the Manual Ability Classification System [MACS] level (range I-V), and the Box and Block Test [BBT].

Range of motion was measured using a goniometer, with the children seated either in a chair or their wheelchair. UERS documentation form (Appendix 1) was used to record children's upper limb passive range of motion of the shoulder, elbow, forearm, wrist, and hand grasp and release[18]. Manual ability was evaluated with MACS, a five-level ordinal system. Level I includes children with minor limitations, while levels IV and V show more severe functional limitations[19]. Gross manual dexterity was assessed with BBT according to Mathiowetz[17,20]. A wooden box, 53.7x25.4 cm, divided into two equal compartments by a 15.2 cm high partition, was prepared. Participants were instructed to transfer as many 2.5 cm cubes as possible from one compartment to the other one at a time in one minute with each hand. Their score was the number of cubes transferred completely.

Statistical analysis

A sample size of 47 patients was required to obtain an expected correlation 0.4, with a two-sided 5% significance level and a power of 80%, in a relevant with a study by Park ES et al[21].

Statistical analysis was performed using GraphPad version 5.0. The strength of association of the data was calculated using Pearson's correlation coefficient for continuous data (UERS and BBT) and Spearman's correlation coefficient for categorical data (UERS and MACS). Both correlation coefficients are to be interpreted as follows: <0.20-very weak; 0.20 to 0.39-weak; 0.40 to 0.59-moderate; 0.60 to 0.79-strong; and 0.80 to 1.0-very strong. Statistical significance was defined as a *p*-value less than 0.05.

Results

The 57 children who participated in the present study included 32 boys and 25 girls. Their ages ranged from 6 to 16 years. The most common types of cerebral palsy were diplegia (40.35%) and spastic (73.68%), according to anatomic and neuromuscular type, respectively. Most of participants were right-handed (52.63%) and had a ambulation status classified as GMFCS V (33.33%). The common upper limb deformities were as follows: forearm pronation (33.33%), finger swan-neck (21.06%), thumb-in-palm (19.30%), wrist flexion (15.78%), shoulder internal rotation (8.77%), and shoulder adduction (3.51%). None had elbow flexion deformity. Other general characteristics of the participants are described in Table 1.

The UERS, MACS and BBT data anlyzed using Pearson's correlation coefficient (UERS and BBT) and Spearman's correlation coefficient (UERS and MACS). The association between UERS and BBT (Figure 1) showed a moderate positive linear relationship ($r = 0.5188$; $p<0.0001$). There was a negative correlation between UERS and MACS (Figure 2), while the Spearman's correlation coefficient indicted a moderate linear relationship ($r = -0.5217$; $p<0.0001$).

Table 1. Patients' general characteristics

Characteristic	Percentage
Gender	
Female	25/57 (43.86%)
Male	32/57 (56.14%)
Type of cerebral palsy (by anatomic)	
Hemiplegia	9/57 (15.79%)
Diplegia	23/57 (40.35%)
Triplegia	6/57 (10.53%)
Quadriplegia	16/57 (28.07%)
Double Hemiplegia	1/57 (1.75%)
Unspecified	2/57 (3.51%)
Type of cerebral palsy (by neuromuscular)	
Spastic	42/57 (73.68%)
Athetoid	5/57 (8.78%)
Ataxia	3/57 (5.26%)
Unspecified	7/57 (12.28%)
Dominant hand	
Right	30/57 (52.63%)
Left	27/57 (47.37%)
GMFCS	
I	7/57 (12.28%)
II	3/57 (5.27%)
III	11/57 (19.30%)
IV	17/57 (29.82%)
V	19/57 (33.33%)
Part of contracture	
Shoulder internal rotation	5/57 (8.77%)
Shoulder adduction	2/57 (3.51%)
Elbow flexion	0/57 (0%)
Forearm pronation	19/57 (33.33%)
Wrist flexion	9/57 (15.78%)
Thumb-in-palm	11/57 (19.30%)
Swan-neck deformity of fingers	12/57 (21.06%)

Discussion

The present study demonstrated a correlation between range of motion represented by UERS and functional performance as determined by MACS and BBT. The correlation between range of motion and MACS was moderate and negative, while a moderate positive correlation was found between UERS and BBT.

It has been reported that upper extremity function of cerebral palsy children is associated with several factors, some of which have been identified as prerequisites for surgical treatment(5). Severity of deformities, especially forearm pronation, wrist flexion, and thumb-in-palm, are significantly related to function of GMFCS, the Upper Limb Physician's Rating Scale [ULPRS], and UERS(4). Hypertonicity of muscle, which is a common finding in cerebral palsy, affects not only functional usage but also pain(22). In addition, it has been reported that patients with spasticity have a better outcome following surgical treatment than those with other neurologic types of cerebral palsy(5). Stereognosis, an ability to perceive and recognize the form of an object in the absence of visual and auditory information, as well as the level of cognition or intelligent quotient also have a positive impact on the outcome of upper extremity treatment. According to a study by Green WT and Banks HH(23), cerebral palsy children who received an operation transferring FCU to ECRB or ECRL had a better post-operative hand and function among groups of patients with intact stereognosis and higher intelligent quotient levels. Hoffer MM et al(24) reported on 38 patients with cerebral palsy who underwent tendon release and transfer. They found that 10 patients who had problems of cognition, sensation, and hand placement failed to gain hand function, while the other 28 patients with better pre-operative criteria had improvement of function.

Range of motion is one of the key components for assessment of extremities and joints. It is a common tool used by physicians which reflects capability for any tasks or activities of daily living(25). Since this

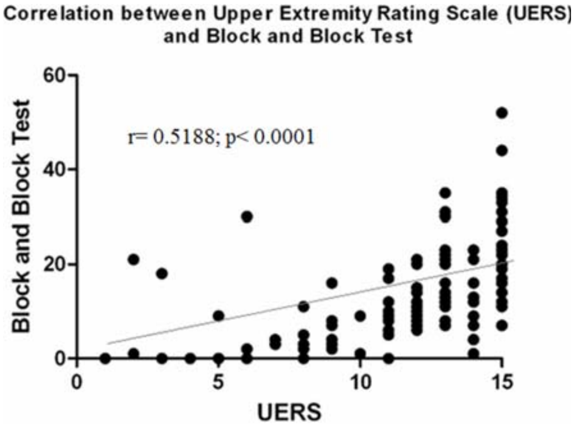

Figure 1. Correlation between UERS and BBT in children with cerebral palsy.

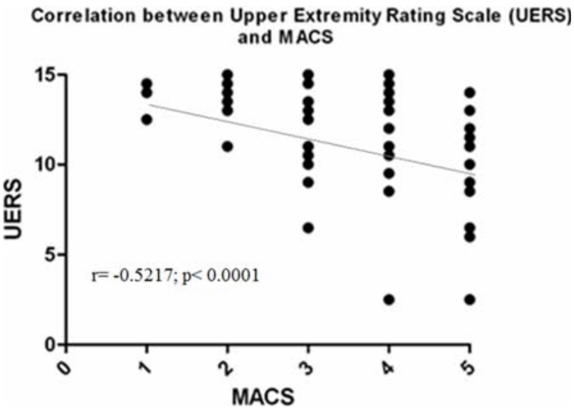

Figure 2. Correlation between UERS and MACS in children with cerebral palsy.

parameter is uncomplicated, it is well suited for evaluating patients with neuromuscular problems such as cerebral palsy. The UERS was designed to provide a score for upper limb active and passive range of motion which is simple, reliable, and reproducible. Koman LA et al[18] found that UERS yielded excellent inter- and intra-rater reliability (interclass correlation coefficients in the range of r = 0.94 to 0.97) and concurrent validity as it was positively correlated to the Melbourne Assessment of Unilateral Upper Limb Function.

The MACS consists of five levels of functional grades which reflect the ability of children's typical usage of their hands for daily activities, but not their maximal capacity. It defines the functionality of the upper extremity of the child as completing any activity with one or both hands or as a bimanual function. However, it is not intended for evaluating each hand separately[16,26]. This classification has a good validity and reliability. It has been reported that the intra-class correlation coefficient between physicians was 0.97 (95% CI 0.96 to 0.98) and that between parents and physicians was 0.96 (95% CI 0.89 to 0.98)[16]. It was designed for children 4 to 18 years of age. This classification is useful not only for physicians but also for parents for several purposes, e.g., social services communication for support and assistance[15,16]. The score is commonly used for evaluation and management in cerebral palsy patients, as it can classify patients using uncomplicated criteria. According to a study by Park ES et al[21], there is a significant relationship between UERS of the non-dominant hand and MACS level in both bilateral and unilateral CP (kappa value of r = -0.71 and r = -0.41, respectively). However, UERS of the dominant side is related to MACS only in bilateral CP, but not in unilateral CP. As in this study, a significant negative correlation between UERS of the non-dominant side and MACS (r = -0.5217; $p<0.0001$) was reported. Since UERS represents range of motion measurements only in the non-dominant hand, these findings cannot be generalized to function of either the dominant hand or to both hands.

The Box and Block test [BBT] is a tool for evaluating unilateral gross manual dexterity, i.e., the ability to make skillful, well-directed arm-hand movements in manipulating fairly large objects in a timed situation[17,27]. The test is simple to perform. In previous studies, it has been shown to be accurate for patients with stroke[28,29]. It has been reported that the test-retest reliability at 6-month intervals rho coefficients were 0.976 and 0.937 for the right and left hands, respectively[30]. Inter-rater reliability of BBT has been validated using Pearson's correlation coefficient, and was found to be very high (r = 1.000 and 0.999 for the right and left hand, respectively)[20]. Golubovic S. et al[31] suggested that manual dexterity is a good predictor of manual abilities and that it can be applied to assess functional performance in cerebral palsy children. The results of this study suggest that the range of upper extremity motion assessed by UERS may represent the child's manual dexterity function with a moderate degree of correlation.

There are some limitations of this study. Firstly, spasticity of muscle, which may affect functional performance, was not assessed. This should be considered as a potential confounding factor. As change in spasticity is dynamic, it is relatively difficult to obtain an accurate assessment. Further studies should include all possible confounding factors which could potentially affect the range of motion in functional performance evaluations. Secondly, this study is a population-based design conducted in a school setting and so represents only one community; results cannot be extrapolated to patients who have a greater degree of CP severity, e.g., hospital-bound patients. Nonetheless, the results from this population base can provide insights into the general group of patients with cerebral palsy. Thirdly, the assessment of MACS was determined only for the more severely affected side which may not be directly related to bimanual performance of the upper extremities. However, the BBT assessed both sides and the relationship between range of motion and BBT was determined for each side individually, thus it indicates the significance of upper extremity motion and gross manual dexterity.

The range of motion of the upper extremities in cerebral palsy is associated with the patients' functional performance and ability to perform daily activities as well as manual dexterity. One of the cornerstones of treatment by health-care professionals is an emphasis on treatment strategies which provide improvement in the range of motion.

What is already known on this topic?

Several factors, including type of cerebral palsy, spasticity severity, degree of intelligence, ambulatory status, etc., are known to be associated with functional performance of upper extremity deformity in cerebral palsy.

What this study adds?

This study demonstrates that the range of motion of upper extremities is significantly associated with MACS and that the range of motion is significantly correlated with BBT and manual dexterity function.

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Potential conflicts of interest

The author declares no conflicts of interest.

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Appendix I. Upper Extremity Rating Scale [UERS]

		Left extremity	Right extremity
A. Shoulder: active motion			
Severe	Abduction 0-30°	☐ [0]	☐ [0]
Moderate	Abduction 31-120°	☐ [1]	☐ [1]
Mild	Abduction 121-160°	☐ [2]	☐ [2]
None	No dynamic deformity (161-180°)	☐ [3]	☐ [3]
B. Elbow: active motion			
Severe	Extension >30°	☐ [0]	☐ [0]
Moderate	Extension 16-30°	☐ [1]	☐ [1]
Mild	Extension 5-15°	☐ [2]	☐ [2]
None	No dynamic deformity (<5°)	☐ [3]	☐ [3]
C. Forearm: active motion (examine with elbow 45–90_)			
Severe	Supination 0-45°	☐ [0]	☐ [0]
Moderate	Supination 46-90°	☐ [1]	☐ [1]
Mild	Supination 91-135°	☐ [2]	☐ [2]
None	Supination 136-180°	☐ [3]	☐ [3]
D. Wrist: active motion (examine with elbow 45–90_)			
Severe	Dorsiflexion 0-45°	☐ [0]	☐ [0]
Moderate	Dorsiflexion 46-70°	☐ [1]	☐ [1]
Mild	Dorsiflexion 71-90°	☐ [2]	☐ [2]
None	Dorsiflexion 91-180°	☐ [3]	☐ [3]
E. Hand			
Ineffective grasp/release (any wrist position)		☐ [0]	☐ [0]
Effective grasp/release (dependent upon wrist position)		☐ [1]	☐ [1]
Effective grasp/release (thumb-independent, 2 or 3 finger pinch)		☐ [2]	☐ [2]
Effective grasp/release (any wrist position)		☐ [3]	☐ [3]