Concurrent Responsiveness of Measures for Assessing Motor-Related Abilities in Individuals with Spinal Muscular Atrophy

Abstract:

Objectives: Spinal muscular atrophy (SMA) is the most common autosomal recessive disease of childhood that affects the spinal cord motor neurons. Individuals with SMA experience diffuse muscle weakness and progressive deterioration of motor function. Follow-up assessment of their changes in motor-related abilities is necessary for understanding the outcomes of intervening clinical trials and physical training programs. A valid measure, the Manual Muscle Test (MMT) is currently used for follow-up assessment of muscle strength in individuals with SMA, while another measure, the Gross Motor Function Measure (GMFM) recently has been recommended as a suitable tool for assessing the individuals’ functional motor skills over time. The purpose of this study was to examine the correlations between change in muscle strength and change in motor function, in order to understand the concurrent responsiveness of MMT and GMFM.

Methods: Fifty-six individuals (mean age=16.02±9.9 years) with type II SMA (the intermediate form) (n=28) or type III SMA (the mildest form) were recruited from a medical centre. All participants had received three assessments of both MMT and GMFM with an interval of two months. The outcome analyzed was coefficient of variation (CV) in scores on MMT and GMFM among the three assessments from each individual. Comparisons were conducted between the CV of GMFM and that of MMT.

Results: The results demonstrated that in the subjects with type II SMA, the CV in MMT scores of the musculature around shoulder, wrist, finger and thumb were significantly correlated with the CV in scores of sitting dimension on GMFM (p<.05). In the subjects with type III SMA, the CV in MMT scores of hip and knee musculature was correlated with the CV in scores of lying and sitting dimensions on GMFM respectively (p<.05). In the multiple regression analysis, the CV in MMT scores of wrist extensors and thumb flexors were important variables for predicting the CV in score of sitting dimension on GMFM in subjects with type II SMA. While for the subjects with type III SMA, CV in MMT scores of hip extensors and knee extensors were the important predictors of the CV in both the scores of lying and sitting dimensions on GMFM.

Conclusion: This study demonstrates evidence of concurrent responsiveness of MMT and GMFM for assessing the individuals with type II or type III SMA three times over a two-month interval. This information can be valuable in view of follow-up assessments when dealing with clients with progressive declining ability such as SMA in clinical setting.