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

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## Introduction

**Spinal muscular atrophy** (SMA) is the most common autosomal recessive disease of childhood that affects the spinal cord motor neurons.[1] Almost all infants with the most severe form (type I SMA) do not survive beyond the first year. Children with the intermediate form (type II SMA) are able to sit but not to walk independently *(Fig.1)*, while in the mildest form (type III SMA) independent ambulation is achieved *(Fig.2)*. The individuals with SMA experience diffuse muscle weakness and progressive deterioration of motor function. Follow-up assessing their change in motor-related abilities are relevantly needed for understanding the outcomes of intervening clinical trials and physical training programs.

A valid measure, the Manual Muscle Test (**MMT**)[2] is currently used for follow-up assessment of muscle strength in individuals with SMA, while another measure, the Gross Motor Function Measure (**GMFM**)[3] recently has been realized as a recommendable 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 for measuring individuals with SMA.

## Methods

Fifty-six individuals (mean age=16.02±9.9 years) with type II SMA (n=28) or type III SMA were recruited from a medical center. All participants had been assessed by using the MMT and GMFM at the same test session. The participants underwent MMT to assess the strength of muscle groups around the limbs' joint, which were the shoulder, elbow, wrist, finger, hip, knee and ankle. By using GMFM, five dimensional gross motor abilities, i.e. lying & rolling, sitting, crawling & kneeling, standing, and walking & running & jumping, were evaluated. Three assessments of MMT and GMFM were carried out in an interval of two months. The outcome analyzed was coefficient of variation (CV) in MMT scores and GMFM scores. A **CV** is a statistical term, which was computed as the standard deviation (SD) of scores among the three assessments divided by the mean of the three assessments within each individual. Correlation analysis and multiple regression analysis of the data were further conducted.

## Results

*The significant findings* (*Table*): In the participants with type II SMA, the CV in MMT scores of musculatures around shoulder, wrist, finger and thumb were significantly correlated with the CV in GMFM score on sitting dimension (p<.05).

***Fig.1*** Type II SMA, the girl could not raise arms above head

***Fig.2*** Type III SMA, the girls was standing up from floor

In the participants with type III SMA, the CV in MMT scores of hip and knee musculatures were respectively correlated with the CV in GMFM score on lying dimension and on sitting dimension (p<.05).

For the multiple regression analysis, in the participants with type II SMA, the CV in MMT scores of wrist extensor and thumb flexor were the significant variables of predicting the CV in GMFM score on the sitting dimension (p<.05).

While in the participants with type III SMA, the CV in MMT scores of hip extensor and knee extensor were the significant predictors of the CV in GMFM score on both the lying and sitting dimensions (p<.05).

## Discussion

A measure's responsiveness is illustrated as a measurement property that allows the assessment of change. Concurrent responsiveness compares score change in overtime on a measure with current performance on another measure. This study demonstrate evidence of concurrent responsiveness of MMT and GMFM for assessing the individuals with type II or type III SMA three times over 2 month intervals. The study findings could be valuable in view of follow-up assessments in clinical situations.

## References

1. Campbell SK, Vander Linden DW, Palisano RJ. Physical Therapy for Children. 3rd ed. Missouri: Saunders; 2006, p.439.
2. Hislop HJ, Montgomery J. Muscle testing techniques of manual examination. WB Saunders Company, Philadelphia, 6th ed., 1995: 12-233.
3. Nelson L, Owens H, Hynan LS, Iannaccone ST; AmSMART Group. The gross motor function measure is a valid and sensitive outcome measure for spinal muscular atrophy. Neuromuscul Disord 2006; 16:374-80.

***Table***. Significant findings of correlation analysis between CV in MMT score and CV in GMFM score for the two type of participants

| | | L't shoulder abduction | L't shoulder adduction | R't shoulder adduction | L't wrist flexion | L't wrist extension | L't finger flexion | L't thumb flexion | L't thumb extension | L't thumb adduction | L't thumb abduction |
|---|---|---|---|---|---|---|---|---|---|---|---|
| **In type II SMA** | *r* | .545(**) | .487(**) | .447(*) | .559(**) | .705(**) | .468(*) | .453(*) | .589(**) | .378(*) | .432(*) |
| **Sitting dimension** | *p* | .003 | .009 | .017 | .002 | .000 | .012 | .016 | .001 | .047 | .022 |
| | | **L't hip flexion** | **R't hip flexion** | **L't hip abduction** | **R't hip abduction** | **L't knee extension** | **R't knee extension** | **L't ankle dorsiflexion** | | | |
| **In type III SMA** | *r* | .419(*) | .405(*) | .493(*) | .584(**) | .661(**) | .743(**) | .391(*) | | | |
| **Lying dimension** | *p* | .027 | .033 | .010 | .001 | .000 | .000 | .044 | | | |
| **In type III SMA** | *r* | | .411(*) | .526(**) | .429(*) | .453(*) | .733(**) | | | | |
| **Sitting dimension** | *p* | | .030 | .006 | .026 | .018 | .000 | | | | |

***r***: **Pearson coefficient;** ***p***: **p-value; L't left side; R't: right side**