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Reliability of the Modified Hammersmith Functional Motor Scale in Young Children with Spinal Muscular Atrophy

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Abstract

Introduction—The test-retest reliability of the Modified Hammersmith Functional Motor Scale (MHFMS) in children with SMA ≤ 30 months (mo) of age was assessed. The age at which typically developing children (TD) achieve maximum MHFMS scores was also explored.

Methods—Twenty-two children with SMA Type II [mean age=20mo; (SD)5mo; range = 9mo–30mo] were tested twice using the MHFMS. Twenty-five TD children [mean age = 18mo; (SD)7mo; range=9mo–30mo] were tested once.

Results—The average difference between MHFMS scores for SMA children was 0.18 [1st assessment mean=12.8(SD 9.8); 2nd test mean=13.0(SD 8.8)]. Reliability was excellent (ICC_{1,3}=0.96, SEM=1.86). TD participants had MHFMS scores ranging from 36 to 40 [mean=39.2(SD1.2)] and achieved maximum test scores at 12 months of age.

Discussion—MHFMS scores in young children with SMA Type II showed excellent test-retest stability. This suggests that the MHFMS can be used reliably in this younger population for clinical trials and follow-up.

Keywords

spinal muscular atrophy; test-retest reliability; Modified Hammersmith Functional Motor Scale; outcome; MHFMS

INTRODUCTION

Spinal muscular atrophy (SMA) is an autosomal recessive disorder characterized clinically by diffuse muscle weakness and genetically by mutation in the *survival motor neuron (SMN)*

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1 gene.¹ Children with SMA have weakness over a wide range of severity. The most rapid rate of functional decline is early in the course, with a progressively slower rate of decline over time; however, some children may demonstrate stable functional status over many years.^{2,3} To establish the effect of any treatment in the natural course of the disease, valid, standardized and reliable evaluation tools are needed.

Functional testing includes the evaluation of activities of daily and motor skill performance and it can provide a measure of potential efficacy of study interventions in clinical trials.⁴ Parents with young children often monitor their child's developmental progression. They appreciate improvements or changes in motor skills more readily than changes in strength or power. Measures to assess motor function in children with SMA Type II and Type III older than 30 mo of age have been reported.^{5–11} However, recent studies suggest that children of younger ages may benefit most from an intervention, since increased age is associated with increased severity of denervation.^{12, 13} Additionally, clinical trials have suggested that children younger than 6 years could be better responders to therapeutic interventions.^{13–16} Currently a limited number of functional outcome measures are available to assess gross motor change in children with SMA ≤ 30 mo of age,¹⁷ and these measures are not disease specific. Since the most active phase of denervation in children with SMA Type II occurs early in the clinical course of the disease, by 3 years of age,¹² it would be ideal to enroll children in clinical trials prior to onset of the chronic phase of denervation, theoretically allowing the greatest therapeutic benefit and improving outcomes.

Obtaining reliable assessments in children with SMA at young ages is uniquely challenging due to inherent issues with behavior and cooperation. In addition, although natural history suggests that gross motor function may plateau early in the disease course for children with SMA and remain stable for periods of time^{18–26}, developmental maturation remains a confounding factor when assessing change in function in younger children. Therefore, in order to better understand the role developmental maturation may play, an important first step is to ascertain the age at which typically developing (TD) children attain the skills on a given outcome measure.

The Modified Hammersmith Functional Motor Scale (MHFMS) was developed based on natural history of the disorder, is a disease-specific scale. It has demonstrated reliability and validity in children with SMA greater than 2 years of age.⁷ It encompasses 20 developmentally acquired gross motor milestones, many of which require behavioral cooperation and participation during testing that is not often seen in younger children. As a child's behavior can affect performance and test score, it is important to assess the reliability of this tool in younger children. Another relevant issue to consider in choosing a motor scale for young children is the age at which TD children achieve success on the scale's items in order to begin to delineate the age at which developmental maturation may no longer influence test scores.

In this study, we assessed the test-retest reliability of the MHFMS in children with SMA Type II who were ≤ 30 mo of age to determine if a currently available disease-specific assessment that has established reliability in older patients could be used reliably in a younger group of patients. An established tool that could measure motor function across a wider age spectrum would have value in both the clinic and clinical trial settings. In addition, we explored the relationship between age and MHFMS test score in TD children ≤ 30 mo of age. This was important to determine the age at which healthy children achieve success on all items on the MHFMS-SMA (≥ 39 points). Previously, Main et al.⁵ reported that the Hammersmith Scale for Children with SMA (HFMS) should be limited to use in those over 30 months, as control children demonstrated wide variability in test scores prior to that age. We hypothesized, based on typical development, that most skills could be

successfully achieved in TD children at an earlier age. For those with SMA, we hypothesized that there would be little change in MHFMS test scores over a 3–6 month period, as we expected minimal influence of developmental maturation and ongoing denervation on functional performance in this short duration based on extensive clinical experience and previous studies in older children.^{7, 11}

MATERIALS AND METHODS

Participants

Children with SMA were evaluated in 2 tertiary care settings in different states as part of a larger multi-center clinical trial network collaboration. Participants with SMA included 22 children ≤ 30 mo of age with SMA Type II (13 females, 9 males) who were participating in a larger natural history study at the University of Utah from September 1, 2001 through January 1, 2008. Children ≤ 30 mo of age who had 2 MHFMS assessments within a 6-month time period were included in this current study. As both the MHFMS and the HFMS demonstrated stability over a 6 month time period in previous studies with older children with SMA, we chose to look at patients over a similar time period.^{7, 11} Participants were excluded if they were receiving any pharmaceutical intervention. Ages at the time of enrollment ranged from 9 to 30 months (mean 20 mo; SD 5 mo). Diagnosis was genetically confirmed by homozygous deletion of *SMN1* and clinically determined by the child's ability to maintain the sitting position when placed. *SMN2* copy number was known in 20 of the SMA children: 2 had 2 copies, 14 had 3 copies, and 4 had 4 copies. All children were in good health with the exception of SMA and did not require BiPAP greater than 12 hours per day.

In addition, 25 healthy TD children ≤ 30 mo of age who had no history of developmental issues (14 females, 11 males) were recruited as a sample of convenience from Chicago and suburbs to examine their performance on the MHFMS. Ages ranged from 9 to 30 months (mean 18 mo; SD 7 mo), a sample size and age range that matched that of the subjects with SMA. All TD children were prescreened by phone prior to enrollment to ascertain that no developmental issues had been previously brought to the attention of the parents by medical professionals, teachers or others. Any children with a history of prenatal or perinatal events with a known risk for developmental delay were excluded. On the day of the assessment, all children were screened for global delay using the Denver Developmental Assessment-II, and no at-risk performance or delays were noted. All enrolled subjects' guardians provided informed consent per Institutional Review Board (IRB) standards at the institution where they participated (University of Utah, Northwestern University).

Instrument and Testing

The MHFMS (Table 1) consists of 20 items, each scored on a 3-point scale based on ability to perform gross motor tasks (2 for unaided, 1 for assistance, 0 for inability). The total test score can range from 0 if the child cannot perform any of the items to 40 if all the items are fully achieved. All items are administered without thoracic or lower extremity orthoses. The test can be completed in 15 to 30 minutes.⁷ Detailed test instructions can be found on www.smaoutcomes.org. All participants with SMA were assessed using the MHFMS administered by the local clinical evaluator twice during a prospective 6 month period.

Assessments were conducted without access to previous results to prevent any duplication or bias. All TD children were assessed once using the MHFMS by a second rater, at their local site. Both raters were physical therapists (PT) with extensive experience (> 250 MHFMS assessments). Reliability in administering and scoring the MHFMS was previously established for both raters [ICC=0.99, 95% CI (0.96, 0.99)].⁷ Several steps were taken to

maximize true responses and minimize variability in our study. For example, to limit rater variability, the same PT administered the MHFMS on both occasions. Additionally, the test was performed in a standardized fashion, and the test environment (location and time of day) was consistent from session to session. Both therapists participated in training sessions to insure standard administration of the test.

Statistical Analyses

The MHFMS is a reliable and valid measure for older children with SMA.⁷ The purpose of this analysis was to establish the test-retest reliability of the MHFMS within young children with SMA; modification of the MHFMS was not an aim of this project. Test-retest reliability measures the ability of a score on the scale to remain constant when there is no assumed change in the property that is being measured.

The distribution of each of the outcome variables was assessed for normality using the Shapiro-Wilk test. Since the data were normally distributed, the intra-class correlation coefficient ($ICC_{1,3}$)²⁷ was utilized to assess test-retest reliability for the group with SMA. In addition, the mean, median and standard deviation of change from baseline (T0) to visit 2 (T1), as well as t-tests were used to assess for change from T0 to T1. The standard error of measure (SEM) was calculated as the standard deviation at T0 multiplied by the square root of 1 minus the reliability. The SEM is used to assess reliable change in the MHFMS.

For the TD group, the age at which participants achieved success on all items (score ≥ 39) was calculated. Descriptive analyses were used to determine items that were most difficult for TD children ≤ 30 mo of age to perform.

RESULTS

MHFMS T0 scores for children with SMA ranged from 1 to 31 (mean=12.8, median=9.5, SD=9.8), and similarly, the T1 scores ranged from 0 to 31 (mean=13.0, median=12.5, SD=8.8) (n=22) (Figure 1). Scores were normally distributed (Shapiro-Wilk, $W=0.926$, $p=0.15$). Test-retest reliability was excellent in the group of children with SMA ≤ 30 mo of age ($ICC_{1,3}=0.96$, SEM=1.86) and similar to that for children 2–12 years of age.⁷

The mean interval between observations for the children with SMA ≤ 30 months of age was 2.9 months (SD=2.0 months, median=3.0 months), with a range of 0.7–6.4 months. A paired t-test indicated the change of 0.18 was not statistically significant at $p=0.79$. This indicates that the test is reliable when administered within 6 months for non-ambulatory children with SMA ≤ 30 mo of age. The SEM was 1.86; this indicates that the MHFMS is sensitive to changes of at least this value.

Total MHFMS scores for TD participants ranged from 36 to 40 (mean=39.2, SD=1.2, median=40.0) (n=25) (Figure 2). Scores were not normally distributed (Shapiro-Wilk $W=0.707$, $p<0.001$) with a high ceiling effect. The age at which TD participants achieved a maximum score (≥ 39) on the test was calculated. A maximum test score (≥ 39) was attained by all TD children ≥ 1 year of age and by 84% (n=21) percent of all TD children in this study. In addition, 40% of children less than 1 year of age (n=2) achieved a maximum test score. Items most difficult for non-affected children under 30 months of age included rolling prone to supine over left and walking greater than 4 steps independently (Table 2).

DISCUSSION

This study demonstrates excellent test-retest reliability of the MHFMS over a 6 month duration in children with SMA Type II ≤ 30 mo of age. The reliability and SEM were

similar to previously established values for children in the 2 to 12 year old age group.⁷ As it can be used reliably in this younger population, the MHFMS may also be useful as a measure to track disease progression in infants and children with SMA within the clinical setting from a very early age. In addition, in contrast to Main et al⁵, we have demonstrated the ability of a small sample of TD children to achieve success on all test items (sum score=39/40) at a very early age (1 year). This suggests that developmental maturation may have less impact on test performance of TD children over 1 year of age than previously suspected.

Although our TD population was small, the age of motor skill acquisition was similar to that published in much larger normative samples.^{28–32} Data from normative samples support skill maturation of many items similar to those on the MHFMS [standing, 4 point kneeling (hands and knees position), crawling, steps] occurring by 12–15 months of age.³⁰ In addition, our TD sample demonstrated success (score of 2) on all MHFMS items by 12 months, which suggests that maturation of skills to achieve a full score on the MHFMS may occur much earlier than that reported by Main and colleagues.⁵ Main et al⁵ previously reported that, in a sample of 30 control infants with no known disabilities, 90% of infants older than 29 months could complete the HFMS scale and score 39/40, with those under 30 months demonstrating wide variability in scores. The small size of both studies' TD samples, as well as potential differences in childrearing practices secondary to cultural differences between our (United States) and Main et al's (United Kingdom) TD population samples, may account for the differing observations. A larger study of TD children ≤ 30 mo of age is warranted to confirm these results.

Irrespective of challenging behaviors in a younger age group, such as resistance, fussiness and inattention, high test-retest reliability was achieved for the children with SMA. Therefore, for functional testing in this study, the presence of typical toddler behaviors did not have a strong influence on reliability, and thus age and behavior were not barriers to reliable testing. However, toddlers vary widely in their degree of comfort and willingness to follow directions in a new setting. Thus, when considering use of the MHFMS in clinical trials with younger children, a period of accommodation to the evaluator may be useful in determining whether a child of this age is an appropriate candidate for enrollment in this type of clinical trial using motor function as the primary outcome. Another consideration in clinical trial design is the use of two baseline visits prior to intervention for children at younger ages. This may increase the infants' familiarity with test procedures and comfort with the examiners administering the tests.

As clinical trials ensue it will be important to have measures that can reliably assess change and performance in those at younger ages. The most active phase of denervation in children with SMA Type II occurs early in the clinical course of the disease^{3, 12}; therefore, it would be ideal to enroll children in clinical trials prior to the irreversible loss of motor neurons, since in theory, earlier administration of treatment may have a better chance of showing effectiveness. However, the availability of measures to reliably assess gross motor change and performance in children with SMA ≤ 30 mo of age has been limited by questions regarding inherent developmental confluence as well as stability of measures over time.

This study supports the use of the MHFMS to assess gross motor change in young children with SMA. In addition, the high test-retest reliability in subjects with SMA, as well as the ability of TD subjects to successfully achieve all MHFMS skills by 12 months of age, suggests that after 12–15 months of age, the MHFMS can be used with minimal risk of developmental confluence. In addition, while developmental maturation has been proposed as a confounding variable in testing young children with SMA, our cohort remained stable over the period of the study, which suggests, as does natural history, that gross motor gains

at this age are minimal compared to age related peers. Further study to assess the sensitivity and responsiveness of the MHFMS to change as well as longitudinal MHFMS data in this younger population, as well as in the SMA population in general, is ongoing.

This study was limited by small sample sizes in the TD and SMA populations, as well as a non-random convenience sample of limited ethnic diversity from one geographic location [TD (Chicago), SMA (Utah)]. A larger study of non-affected TD and affected children ≤ 30 mo of age from different ethnic backgrounds and geographical locations is warranted to confirm these results. Others have reported differences in attaining gross motor skills among ethnic groups with varied child rearing practices.^{36–40}

In conclusion, natural history data in SMA supports a progressive loss of functional motor units with increasing age, as well as an increased burden of secondary complications, including contractures and scoliosis. Thus, the expected benefits of any therapy in children with SMA Type II may be greater if children are treated as early as possible following their initial diagnosis. We have demonstrated that the MHFMS can be reliably used in children with SMA younger than 30 mo of age in a clinical trial setting. We hope that this data will encourage others to more readily use this outcome measure in both the clinical and research settings to facilitate collection of a wider dataset in a younger cohort of children with SMA Type II.

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Abbreviations

T0	Baseline visit
HFMS	Hammersmith Functional Motor Scale
ICC	Intra-class correlation coefficient
MHFMS	Modified Hammersmith Functional Motor Scale
mo	Months
SMA	Spinal muscular atrophy
SD	Standard deviation
SEM	Standard error of measure
SMN	Survival motor neuron
TD	Typically developing
T1	Visit 2

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Figure 1. Bivariate Fit of Score of first visit by score from second visit in SMA Type II patients ≤ 30 months.

Figure 2. Modified Hammersmith Functional Motor Scale scores in typically developing children. Scores ≥ 39 indicate that subjects were able to perform all test items.

Table 1

List of Modified Hammersmith Functional Motor Scale test items

Item Description
Frog/chair sitting- no hand support
Long sit- no hand support
Raises one hand to ear level (right/left) (in sitting)
Raises 2 hands to ear level (in sitting)
Gets from sitting to lying (safely, not accidentally)
Lifts head from surface in supine
½ roll from supine, both ways
Rolls supine to prone over right
Rolls prone to supine over left
Rolls supine to prone over left
Rolls prone to supine over right
Lifts head from prone (arms down by sides)
Achieves prop on forearms- head up
Achieves prop on extended arms- head up
Achieves hands and knees
Crawls on hands and knees
Gets to sitting from lying through side-lying
Stands holding on with 1 hand
Stands independently: Count > 3
Takes > 4 steps independently

Table 2

Five test items that were more difficult for the typically developing participants that did not achieve a full score on the Modified Hammersmith Functional Motor Scale

TEST ITEM	Percentage (and number) of total TD children tested unable to achieve maximum score of 2	Percentage (and number) of total TD children tested unable to achieve partial score of 1
Takes > 4 steps independently	20% (4/25) *	0%
Rolls prone to supine over left	16% (4/25)	0%
Lifts head in supine	12% (3/25)	4% (1/25) [‡]
Stands independently	12% (3/25) [†]	0%
Rolls prone to supine over right	4% (1/25)	0%

TD, typically developing.

* The 4 youngest subjects were unable to take > 4 steps, but all scored a 1 on this item by taking at least 2–4 steps unaided.

[†] The 3 youngest subjects were unable to stand independently for ≥ 3 seconds, but all stood independently for < 3 seconds.

[‡] All children were able to achieve at least a partial score of 1 on all test items with the exception of the youngest child at 9 months who was unable to lift or attempt to lift his head from supine.