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Modified Hammersmith Functional Motor Scale for Children with Spinal Muscular Atrophy
(MHFMS-SMA / MHFMS-Extend)

Availability:	<p>For more information about Modified Hammersmith Functional Motor Scale and the MHFMS-EXTEND modules, visit Project Cure SMA - Outcome Measures</p> <p>For questions related to content and test use, please contact Kristin Krosschell, PT, MA, PCS: k-krosschell@northwestern.edu</p>
Classification:	<p>Supplemental – Highly Recommended for Mitochondrial Disease (Mito) as a primary outcome measure in treatment trials of young non-ambulatory children with SMA.</p> <p>Supplemental – Highly Recommended for Congenital Muscular Dystrophy (CMD)</p> <ul style="list-style-type: none"> • Highly recommended for studies analyzing motor function - age limit 2+
Short Description of Instrument:	<p>The Modified Hammersmith Functional Motor Scale for Children with Spinal Muscular Atrophy (MHFMS-SMA) (Krosschell, et al 2006) is an adaptation of the original Hammersmith Functional Motor Scale for Children with SMA (Main et al, 2003). The original Hammersmith Functional Motor Scale for Children with SMA was modified to establish a standard measure of functional ability in children with non-ambulant spinal muscular atrophy Types II and III for use in longitudinal multi-center clinical trials. Concrete operational definitions were developed and scoring criteria were clarified to minimize potential ambiguities in the administration and scoring of the test. In addition, after a pilot study, item order of 20 motor tasks was modified to minimize position changes and to decrease fatigue and undue stress on the children during testing (Krosschell et al, 2006). The MHFMS is currently in use to assess gross motor abilities of non-ambulant children with SMA in multiple research trials as well as in clinical settings.</p> <p>The MHFMS-Extend was later developed to address ceiling effects in stronger ambulant children with SMA. The MHFMS-Extend consists of the MHFMS and an extended gross motor module and was developed to allow for assessment of stronger children with SMA Type II and for assessment of ambulant children with SMA Type III. Items selected for the MHFMS-Extend were chosen based on functional, clinical and research applicability after careful assessment of a large group of children with SMA Type III and a group of stronger Type II children. Additional test modules include a Timed Test Module and a Fine Motor Module. The test manuals for each were developed to assure standardized assessment in clinical trials and to allow for training of new trial evaluators and clinicians working with children with SMA.</p>

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Scoring Information:	The Hammersmith Scale is scored on a Likert scale. Each item is scored as 0 (unable), 1 (partially completes), or 2 (completes independently). A total score is calculated ranging from 0-40, with 40 being the highest score. No sub-score is calculated.
Time to Administer:	10 to 15 minutes with a cooperative child
Comments/Special instructions:	<p>Strengths/Weaknesses: The Hammersmith functional motor scale for children with SMA is intended to be sensitive to those functional motor deficits of children with SMA that result directly from weakness. Hammersmith functional motor scale for children with SMA has been shown to be both quick to administer and reliable in non-ambulant children with SMA as young as 30 months of age [Main et al 2003], when other tools such as myometry or the medical research council (MRC) scale cannot be easily or reliably performed.</p> <p>The Hammersmith functional motor scale for children with spinal muscular atrophy was modified to establish a standard measure of functional ability in children with non-ambulant spinal muscular atrophy types 2 and 3 in a longitudinal multi-center clinical trial. In children under 5 years of age, functional motor scales may be more appropriate than myometry in the assessment of strength in children with SMA, as motivation for maximum performance need not depend upon comprehension of the purpose of the task. Scales targeted to assess functional ability with disease-specific and strength-specific tasks may have the potential to exceed direct measures of power in sensitivity and reliability (Main et al 2003, Chung et al 2004, Iannaccone 2002). Functional testing may not be able to detect subtle changes and/or monitor for changes in muscle strength (Moxley 1990). However, functional tests are better at assessing outcomes (activities of daily living) that may be more meaningful to patients (Krosschell et al 2006).</p> <p>Krosschell et al (2006) assessed the intra- and interrater reliability and the test-retest stability of a modified version of the scale. Both intra- and interrater reliability were established. Intrarater reliability between the first and second test had a reliability coefficient of 0.99 indicating excellent reliability within a given rater. Interrater reliability score and rescore reliability assessed by intraclass correlation coefficient demonstrated an interrater reliability of 0.953 at a 95 % confidence interval (0.913, 0.982). Results indicated that the scale is reliable and stable over a 6 month period. Reliability was maintained when patient sample criteria were expanded to include children younger than 30 months and children with popliteal angles greater than 20 degrees .</p>

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<p>Comments/Special instructions continued:</p>	<p>These data established the modified Hammersmith functional motor scale for children with spinal muscular atrophy as a reliable instrument for use in multi-center treatment trials in non-ambulant spinal muscular atrophy children. Data from this study provided additional support for the use of original scale items in terms of ease of administration, usefulness and reliability, while incorporating modifications to optimize its use in a multi-center clinical research setting. The outcome measures were sensitive to small changes in functional ability. The modified Hammersmith functional motor scale appeared to be well suited for use as a primary outcome measure in treatment trials of young non-ambulatory children with SMA who are able to sit unsupported, although it could be used in weaker children unable to sit or in stronger SMA children who have the ability to stand or even take a limited number of steps unsupported, though less ideal. This tool was reliable in children as young as 2 years of age.</p> <p>Psychometric Properties: Items selected for the MHFMS-Extend were chosen based on functional, clinical and research applicability after careful assessment of a large group of children with SMA Type III and a group of stronger Type II children. Additional test modules include a Timed Test Module and a Fine Motor Module which could be considered as add-on modules. The test manuals for each were developed to assure standardized assessment in clinical trials and to allow for training of new trial evaluators and clinicians working with children with SMA.</p> <p>Administration: Should be administered by expert clinicians.</p>
<p>Rationale for Recommendations:</p>	<p>This tool has potential applications for young children with mitochondrial myopathies and marked weakness, particularly those with mtDNA depletion myopathy secondary to TK2 deficiency which has an SMA phenotype and may present in infancy.</p> <p>Validated in SMA pediatric patients. Not yet validated in Mito? Children (2 to 11 Years) Adolescents (12 to 15 Years)</p>

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References:	<p>Krosschell KJ1, Maczulski JA, Crawford TO, Scott C, Swoboda KJ. A modified Hammersmith functional motor scale for use in multi-center research on spinal muscular atrophy. <i>Neuromuscul Disord.</i> 2006 Jul;16(7):417-26.</p> <p>Chung BH, Wong VC, Ip P. Spinal muscular atrophy: survival pattern and functional status. <i>Pediatrics.</i> 2004;114(5):e548–e553. [PubMed]</p> <p>Iannaccone ST. Outcome measures for pediatric spinal muscular atrophy. <i>Arch Neurol.</i> 2002;59(9):1445–1450. [PubMed]</p> <p>Main M, Kairon H, Mercuri E, Muntoni F. The Hammersmith functional motor scale for children with spinal muscular atrophy: a scale to test ability and monitor progress in children with limited ambulation. <i>Eur J Paediatr Neurol.</i> 2003;7(4):155–159. [PubMed]</p> <p>Moxley RT., 3rd Functional testing. <i>Muscle Nerve.</i> 1990;13:S26–S29. [PubMed]</p>
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