

**NINDS CDE Notice of Copyright
Myotonic Dystrophy Health Index (MDHI)**

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| Availability: | <p>Author: Chad Heatwole (chad_heatwole@urmc.rochester.edu) The Neuromuscular Institute of Quality-of-Life Studies and Outcome Measure Development</p> <p>Licensing opportunities for the MDHI instrument are available through the Office of Technology Transfer (OTT) at the University of Rochester Medical Center (URMC) - University of Rochester Office of Technology Transfer (Search for technology number 6-2189).</p> <p>Please visit these websites for more information about the instrument:</p> <ul style="list-style-type: none"> • The Myotonic Dystrophy Health Index (MDHI) • The Myotonic Dystrophy Health Index (MDHI) |
| Classification: | <p>Supplemental-Highly Recommended: Myotonic Dystrophy (DM)</p> <ul style="list-style-type: none"> • Highly recommended for use as a primary or secondary outcome measure during myotonic dystrophy clinical trials. • Highly recommended as a marker of disease severity for patients with myotonic dystrophy. |
| Short Description of Instrument: | <p>The MDHI is a disease-specific patient reported outcome measure specifically created for myotonic dystrophy type-1 (DM1). It was designed to measure a patient's health state and to be used as a responsive outcome measure during clinical trials. It measures a patient's perception of their total health and 17 areas of sub-health. Subscales include: mobility, upper extremity function, ability to do activities, fatigue, pain, gastrointestinal issues, vision, communication, sleep, emotional issues, cognitive impairment, social satisfaction, social performance, myotonia, breathing, swallowing, and hearing.</p> |
| Scoring: | <p>The total MDHI score and a score for each subscale is produced each time a patient completes the instrument. Scores range from 0 to 100 with 100 representing the most severe disease. Standard values are available for the MDHI total score and each subscale score based on gender, age, CTG repeat length, education level, and employment status.</p> <p>Administration time is 15-20 minutes and does not require supervision by clinical staff.</p> |

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| Rationale for Recommendations: | <p>The MDHI was developed for use specifically for the DM1 population. It was validated using standard techniques and a large population of DM1 patients.</p> <p>The MDHI has been tested for validity and reliability in DM1. 278 DM1 patients identified the most relevant questions for the MDHI. After factor analysis, patient interviews, and test-retest reliability assessments was done to refine and evaluate this instrument. Questions in the final MDHI represent 17 areas of DM1 health. The internal consistency was acceptable in all subscales. The MDHI had a high test-retest reliability (ICC = 0.95) and differentiated between DM1 patient groups with different disease severities. Initial evaluation of the MDHI provides evidence that it is valid and reliable as an outcome measure for assessing patient-reported health. These results suggest that important aspects of DM1 health may be measured effectively using the MDHI. In addition this instrument is now used in multiple clinical trials including the ongoing DM1 EU-study OPTIMISTIC.</p> |
| References: | <p>Heatwole, C, Bode, R, Johnson, N, Dekdebrun, J; Dilek, N; Heatwole, M; Hilbert, J, Luebbe, E; Martens, W; McDermott, M; Rothrock, N; Thornton, C; Vickrey, B; Victorson, D; Moxley, R. The Myotonic Dystrophy Health Index: Initial Evaluation of a New Outcome Measure. Muscle and Nerve. 2013.</p> <p>Heatwole C, Bode R, Johnson N, et al. Patient-reported impact of symptoms in myotonic dystrophy type 1 (PRISM-1). Neurology 2012;79:348-57.</p> |