

NINDS CDE Notice of Copyright
The Newcastle Mitochondrial Disease Adult Scale

Availability:	Freely available: The Newcastle Mitochondrial Disease Adult Scale Link
Classification:	Supplemental: Mitochondrial Disease
Short Description of Instrument:	<p>The NMDAS has been introduced to allow evaluation of the progression of mitochondrial disease in adult patients over 16 years. The Newcastle Mitochondrial Disease Paediatric Scale (NMDPS) provides a similar assessment tool for paediatric patients. Repeated administration of the scale provides a quantitative assessment in the longitudinal follow up of patients with mitochondrial disease of any genetic cause. The use of this rating scale will standardise patient assessment and ensure more accurate data collection to aid our understanding of the natural history of mitochondrial disease. It is predicted that the scale will also prove to be an invaluable tool for future clinical assessment of proposed treatments.</p> <p>The rating scale encompasses all aspects of mitochondrial disease by exploring several domains: Current Function; System Specific Involvement; Current Clinical Assessment and Quality of Life.</p>
Scoring Information	<p>Scoring: Each question in the NMDAS has a possible score from 0-5. Each of the first 3 section scores are calculated by simply summing the scores obtained for each question in that section. The higher the score the more severe the disease.</p> <p>The Quality of Life Section undergoes separate scoring as detailed in the SF manual. It is helpful to present the scores for each section rather than simply referring to an overall score for the whole scale. At the very least, it is imperative that the quality of life score obtained on the SF-12v2 is presented separately from the disease score obtained in sections I-III.</p>
Comments/Special Instructions:	<p>This assessment scale appears to have been created specifically as a tool to follow disease progression. It is broad, possibly too broad, but has been validated. It is designed to cover the systemic nature of mitochondrial disease so does not go into the same detail as do some of the more specific tools (e.g. SARA cerebellar scales). It includes quality of life questions and self-reported psychiatric evaluation.</p> <p>There is a detailed manual available with instructions on how to score and it is clear that strict adherence to the rules for scoring is required if the tool is to function properly. This scale was developed by and for neurologists, and is probably best suited for diseases in which neurological dysfunction is an integral part. It does however, cover all systems.</p> <p>Limitations: No obvious limitations. The scoring requires normal level of neurological competence to perform some of the clinical evaluations.</p> <p>Strengths/Advantages: Appears robust for following multisystemic diseases that are typical of mitochondrial dysfunction.</p>

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<p>Rationale / Justification:</p>	<p>Psychometric Properties: This rating scale has been validated by the Newcastle Mitochondrial Disease Research Group on two separate occasions through the administration of the scale by 4 clinicians to 16 and 15 patients respectively. The scale demonstrated good to excellent agreement between raters for individual domain scores and the overall disease score. However, this agreement can only be ensured if all users of the scale adhere closely to the instructions given in this manual. Individual interpretation of the questions will alter the scores assigned and consequently reduce the consistency and reliability of the data collected.</p> <p>Administration: We advise that the scale should be administered at either six or twelve month intervals by clinicians with experience in the care of patients with mitochondrial disease.</p>
<p>Population / Age Range / Validation:</p>	<p>Patients >16 years of age.</p> <p>It appears possible to perform assessment in patients who are not themselves capable of providing information i.e. through a caregiver. The tool appears best suited to patients in whom disease affects several systems and progression in one or more can vary.</p> <p>This instrument can be applied to all types of mitochondrial disease. Single organ diseases, such as LHON, may be least suited, but even here the scale covers the essential requirements for following disease progression. Neurological diseases are probably best suited.</p> <p>It has been validated in MELAS, single deletion disorders amongst others.</p>

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References:	<p>Grady JP, Campbell G, Ratnaïke T, Blakely EL, Falkous G, Nesbitt V, Schaefer AM, McNally RJ, Gorman GS, Taylor RW, Turnbull DM, McFarland R. Disease progression in patients with single, large-scale mitochondrial DNA deletions. <i>Brain</i>. 2014 Feb;137(Pt 2):323-34. ⁱ</p> <p>Nesbitt V, Pitceathly RD, Turnbull DM, Taylor RW, Sweeney MG, Mudanohwo EE, Rahman S, Hanna MG, McFarland R. The UK MRC Mitochondrial Disease Patient Cohort Study: clinical phenotypes associated with the m.3243A>G mutation—implications for diagnosis and management. <i>J Neurol Neurosurg Psychiatry</i>. 2013;84(8):936-8. ⁱⁱ</p> <p>de Laat P, Smeitink JA, Janssen MC, Keunen JE, Boon CJ. Mitochondrial retinal dystrophy associated with the m.3243A>G mutation. <i>Ophthalmology</i>. 2013Dec;120(12):2684-96.</p> <p>Bates MG, Hollingsworth KG, Newman JH, Jakovljevic DG, Blamire AM, MacGowan GA, Keavney BD, Chinnery PF, Turnbull DM, Taylor RW, Trenell MI, Gorman GS. Concentric hypertrophic remodelling and subendocardial dysfunction in mitochondrial DNA point mutation carriers. <i>Eur Heart J Cardiovasc Imaging</i>. 2013 Jul;14(7):650-8.</p> <p>de Laat P, Koene S, van den Heuvel LP, Rodenburg RJ, Janssen MC, Smeitink JA. Clinical features and heteroplasmy in blood, urine and saliva in 34 Dutch families carrying the m.3243A > G mutation. <i>J Inherit Metab Dis</i>. 2012;35(6):1059-69.</p> <p>Mancuso M, Orsucci D, Ienco EC, Pini E, Choub A, Siciliano G. Psychiatric involvement in adult patients with mitochondrial disease. <i>Neurol Sci</i>. 2013;34(1):71-4.</p> <p>Newcastle Mitochondrial Disease Adult Scale</p> <p>Newcastle Mitochondrial Disease Adult Scale Manual</p>
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ⁱ Authors of the instrument and use of this scoring system as used in a study are cited here.

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