

Upper limb functional assessment scale for children with Duchenne muscular dystrophy and Spinal muscular atrophy

Escala de evaluación funcional de extremidades superiores en niños con distrofia muscular de Duchenne y Atrofia músculo espinal

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Abstract

Introduction: Duchenne muscular dystrophy (DMD) and Spinal muscular atrophy (SMA) causes significant disability and progressive functional impairment. Readily available instruments that assess functionality, especially in advanced stages of the disease, are required to monitor the progress of the disease and the impact of therapeutic interventions. **Objective:** To describe the development of a scale to evaluate upper limb function (UL) in patients with DMD and SMA, and describe its validation process, which includes self-training for evaluators. **Patients and Method:** The development of the scale included a review of published scales, an exploratory application of a pilot scale in healthy children and those with DMD, self-training of evaluators in applying the scale using a handbook and video tutorial, and assessment of a group of children with DMD and SMA using the final scale. Reliability was assessed using Cronbach and Kendall concordance and with intra and inter-rater test-retest, and validity with concordance and factorial analysis. **Results:** A high level of reliability was observed, with high internal consistency (Cronbach $\alpha = 0.97$), and inter-rater (Kendall $W = 0.96$) and intra-rater concordance ($r = 0.97$ to 0.99). The validity was demonstrated by the absence of significant differences between results by different evaluators with an expert evaluator ($F = 0.023$, $P > .5$), and by the factor analysis that showed that four factors account for 85.44% of total variance. **Conclusions:** This scale is a reliable and valid tool for assessing UL functionality in children with DMD and SMA. It is also easily implementable due to the possibility of self-training and the use of simple and inexpensive materials.

Keywords:

Neuromuscular diseases; Scale; Function; Duchenne muscular dystrophy; Spinal muscular atrophy; Upper limb.

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