

A motor function measure scale for neuromuscular diseases; validation study

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In order to improve the knowledge of natural history of each neuromuscular disease, to select patients for therapeutic trials and to quantify outcomes of therapeutic measures, precise tools are required to objective motor disabilities of patients.

Objective: validation of the Motor Function Measure, a clinical scale designed for neuromuscular diseases of children and adults.

The validation study took place in 19 units and included 303 patients, aged 6 to 62 years. 72 patients had Duchene muscular dystrophy, 32 Becker muscular dystrophy, 30 limb-girdle muscular dystrophy, 39 facio-scapulo-humeral dystrophy, 29 myotonic dystrophy, 21 congenital dystrophy, 10 congenital muscular dystrophy, 35 spinal muscular atrophy and 35 hereditary neuropathy. The scale comprised 32 items, with ratings from 0=no movement to 3 = complete movement, testing a variety of movements (in lying, standing and sitting positions). The sensitivity to change was tested with 152 patients.

A factor analysis confirmed the structure of the scale with 3 dimensions : standing position and transfers, axial and proximal motor function, distal motor function. Agreement coefficients for inter-rater reliability were excellent for 9 items, good for 20 items and moderate for 3 items. High correlations were found between the total score and Vignos ($r=0.91$) and Brooke ($r=0.85$) grades, Functional Independence Measure ($r=0.88$) and physiotherapists ($r=0.91$). A subsample of patients was evaluated one year later.

The Duchene patients ($N=41$) showed the highest decrease: 5.5 ± 6.1 ($p<0.0001$). The effect size (SRM) in this group is 0.95.

This scale is reliable, does not require any special equipment and is well accepted by patients. Its sensitivity to change enables its use in clinical trials of neuromuscular diseases, especially in DMD patients.