A motor function measure for neuromuscular diseases. Construction and validation study.

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Abstract
A new scale for motor function measurement has been developed for neuromuscular diseases. The validation study included 303 patients, aged 6-62 years. Seventy-two patients had Duchenne muscular dystrophy, 32 Becker muscular dystrophy, 30 limb-girdle muscular dystrophy, 39 facioscapulo-humeral dystrophy, 29 myotonic dystrophy, 21 congenital myopathy, 10 congenital muscular dystrophy, 35 spinal muscular atrophy and 35 hereditary neuropathy. The scale comprised 32 items, in three dimensions: standing position and transfers, axial and proximal motor function, distal motor function. Agreement coefficients for inter-rater reliability were excellent (kappa=0.81-0.94) for nine items, good (kappa=0.61-0.80) for 20 items and moderate (kappa=0.51-0.60) for three items. High correlations were found between the total score and other scores: Vignos (r=0.91) and Brooke (r=0.85) grades, Functional Independence Measure (r=0.91), the global severity of disability evaluated with visual analog scales by physicians (r=0.88) and physiotherapists (r=0.91). This scale is reliable, does not require any special equipment and is well-accepted by patients. Its sensitivity to change is being assessed to permit its use in clinical trials of neuromuscular diseases.

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