



**The Amyotrophic Lateral Sclerosis Functional Rating Scale. Assessment of activities of daily living in patients with amyotrophic lateral sclerosis. The ALS CNTF treatment study (ACTS) phase I-II Study Group.**

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**OBJECTIVES:** To test the utility of a new, easy to administer instrument for assessing activities of daily living in patients with amyotrophic lateral sclerosis (ALS), to validate its accuracy, and to assess its ability to record disease progression in patients with ALS against other functional scales, quantitative isometric muscle testing, and global assessment scales. **DESIGN:** Serial assessments of patients who presented to four ALS treatment centers in two multicenter studies. **PATIENTS:** Study 1 (cross-sectional) evaluated 75 consecutive patients who presented to four ALS treatment centers during a 2-month period. Study 2 (longitudinal) evaluated the progression of 53 patients who were enrolled in a multicenter, phase I-II clinical trial of recombinant human ciliary neurotrophic factor for treatment of ALS. **OUTCOME MEASURES:** The ALS Functional Rating Scale (ALSFRS) was compared with quantitative myometry and with other measures of daily function in patients with ALS both cross-sectionally and longitudinally. **RESULTS:** The first study of 75 patients evaluated the internal consistency, the test-retest reliability, and the construct validity of the ALSFRS. Internal consistency and test-retest reliability were high. Patient self-rating of upper- and lower-extremity-dependent tasks were highly correlated with measures of upper- and lower-extremity strength, respectively. Thus, the ALSFRS has good construct validity. In the second study, ALSFRS scores declined in tandem with deterioration in motor and pulmonary function, indicating its sensitivity to change. **CONCLUSIONS:** The ALSFRS is a useful instrument for evaluation of functional status and functional change in patients with ALS. Its results are in close agreement with objective measures of muscle strength and pulmonary function. The ALSFRS may be used as a screening measure for entry into clinical trials, as a surrogate measure of function in situations in which muscle strength cannot be measured directly, or as an adjunct to myometry.