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Appraisal

Clinimetrics: Amyotrophic Lateral Sclerosis Functional Rating Scale-revised (ALSFRS-R)

Summary

Description: The Amyotrophic Lateral Sclerosis Functional Rating Scale-revised (ALSFRS-R) is a self-reported questionnaire that measures physical function in patients with amyotrophic lateral sclerosis.[1] The ALSFRS-R is the most commonly used functional rating instrument in clinical practice and clinical trials. In amyotrophic lateral sclerosis, reduced physical function and the rate of decline are used as markers of disease severity and disease progression. The ALSFRS-R is easy to use and can be administered within 10 minutes. Furthermore, ALSFRS-R has been adapted for online,[2] telephone,[3,4] and self-administration,[5] and is translated into several languages.[6–9]

Instructions for completing and scoring: The ALSFRS-R is a 12-item rating scale across four functional domains: fine motor, gross motor, bulbar and respiratory function. Each item consists of five possible responses (0 = unable to perform the task to 4 = normal functioning). The 12 items address daily living activities: speech, salivation, swallowing, handwriting, preparing food, dressing, bed mobility, walking, climbing stairs, dyspnoea, orthopnoea, and breathing. Conventionally, individual item scores are added to yield a combined score between 0 and 48 (maximum score). However, due to the heterogeneity and multidimensionality of amyotrophic lateral sclerosis, it is recommended that domain-specific subscores be reported, as they provide better prognostic value and more accurately reflect disease severity.[10,11] In fact, univariate analysis has identified that an ALSFRS-R respiratory subscore of < 10 at initial assessment is an adverse prognostic marker in amyotrophic lateral sclerosis.[12]

The rate of change of the ALSFRS-R score (termed the 'ALSFRS-R slope') has also been shown to be an acceptable prognostic indicator.[12–14] The total ALSFRS-R slope can be calculated using the formula:

$$\frac{48 - (\text{Total ALSFRS-R score at initial assessment})}{\text{time from onset to initial assessment } (\textit{months})}$$

In addition, subscore slopes can be calculated by subtracting each subscore from the maximum subscore possible in each functional domain (eg, 12 for bulbar and respiratory, and 24 for motor combined) divided by symptom duration. Assessment of specific subscore slopes is important when considering the differing rates of decline between bulbar-onset and spinal-onset patients.[11]

Prognosis and survival: The ALSFRS-R slope has been identified as an independent predictor of survival (hazard ratio (HR) = 2.8)[13] and a sensitive prognostic biomarker.[12] A higher ALSFRS-R slope score ($\geq$ 1point/month) indicates faster functional decline and is associated with shorter survival.[12,13] More specifically, a slope of < 0.47 at initial assessment is predictive of a longer survival (median survival of 2.4 years, HR = 1), compared with a slope score between 0.47 and 1.11 (median survival 1.6 years, HR = 1.77), or a slope score that is > 1.11, which has the poorest survival (median survival 0.7 years, HR = 3.74).[12]

Reliability and validity: The ALSFRS-R has demonstrated good to excellent inter-rater (ICC = 0.87 to 0.97), intra-rater (ICC = 0.93 to 0.97), and test-retest reliability (ICC = 0.975, r = 0.85 to 0.91).[1,3,5–9,15] The 12-item subscale shows good internal consistency (Cronbach's alpha = 0.71 to 0.93)[1,7,8] but the sum of items to a total score shows weak factorial validity and poor unidimensionality.[16–18] Reporting subscores to provide a disease profile instead of a single combined score is strongly recommended.[10,11] Furthermore, there are contrasting views about whether four or three subscales exist by combining fine and gross motor function into a single 'motor' domain. Rasch analysis of the ALSFRS-R has identified three domains,[17,18] although an exploratory factor analysis indicates that four domains with two cross-loading items (bed mobility, dressing) is superior.[10]

Commentary

Current evidence suggests that the ALSFRS-R constitutes a profile of functional domain subscores rather than a single (total) score representing disease severity, and the slope of ALSFRS-R has important prognostic value. The popularity of the ALSFRS-R is likely due to its ease of use, minimal training required, low cost, and shortage of other recognised biomarkers to quantify disease progression. Further development of the ALSFRS-R and its subscales is required to ensure adequate measurement properties,[10,17] and future research should determine sensitivity to treatment outcomes.

Provenance: Invited. Not peer reviewed.

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