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The ALSFRS-R: a revised ALS functional rating scale that incorporates assessments of respiratory function. BDNF ALS Study Group (Phase III).

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The ALS Functional Rating Scale (ALSFRS) is a validated rating instrument for monitoring the progression of disability in patients with amyotrophic lateral sclerosis (ALS). One weakness of the ALSFRS as originally designed was that it granted disproportionate weighting to limb and bulbar, as compared to respiratory, dysfunction. We have now validated a revised version of the ALSFRS, which incorporates additional assessments of dyspnea, orthopnea, and the need for ventilatory support. The Revised ALSFRS (ALSFRS-R) retains the properties of the original scale and shows strong internal consistency and construct validity. ALSFRS-R scores correlate significantly with quality of life as measured by the Sickness Impact Profile, indicating that the quality of function is a strong determinant of quality of life in ALS.