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Title: Factors predicting survival in amyotrophic lateral sclerosis patients on non-invasive ventilation

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Body: AIM: to determine prognostic factors of survival in patients with amyotrophic lateral sclerosis (ALS) who initiated non-invasive ventilation (NIV). METHODS: retrospective analysis (1999-2012). We included all patients with diagnosis of ALS treated with NIV. Data were collected at initiation of NIV. Variables collected included: sex, age, site of disease onset, time from first symptoms to diagnosis, time from diagnosis to NIV, Forced Vital Capacity percentage (FVC%), PaO2, PaCO2, bulbar involvement at initiation of NIV, NIV compliance. We determined the median survival from initiation of NIV and factors predicting survival. Statistical analysis was performed using the Kaplan-Meier test and Cox proportional hazard models. RESULTS: 175 patients were included; 60% men, age 64 (9) years. 72% of the cases the disease onset was limb. PaO2 76.73 (12) mmHg, PaCO2 45.9 (9) mmHg, FVC 52.9% (16), mean NIV compliance 9.6 h/day. Mean time form first symptoms to diagnosis 11.7 months. 34% of patients had a moderate-severe bulbar involvement at initiation of NIV. Median survival was 14.9 months (mean 19.5 months). In univariate analysis variables associated with survival were delay in diagnosis (HR 0.977, p 0.018), bulbar involvement at initiation of NIV (HR 1.92, p 0.000) and FVC% (HR 0.987, p0.048). Site of onset tended to be associated with survival, without reaching the statistical significance. In multivariate analysis the only covariate that had significant independent prognostic value was bulbar involvement (HR 1.78, p 0.0004). CONCLUSIONS: In our study moderate-severe bulbar involvement at initiation of NIV was the main prognostic factor independent for survival in ALS patients after NIV.