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Title: Predictive factors of survival in amyotrophic lateral sclerosis patients with respiratory dysfunction

Dr. Inês 20919 Neves inesneves.porto@gmail.com MD [1], Dr. Hans 20920 Dabo hansdabo@yahoo.com.br MD [1], Dr. Inês 20921 Belchior inesbelchior@gmail.com MD [1], Dr. Anabela 20922 Marinho a.n.marinho@tvtel.pt MD [1], Dr. Tiago 20923 Pinto tiagoffpinto@gmail.com [1], Prof. Miguel 20925 Gonçalves goncalvesmr@gmail.com [1,2] and Prof. Dr João Carlos 20934 Winck jwinck@hsjoao.min-saude.pt MD [1,2]. [1] Pneumology Department, Centro Hospitalar de São João, Porto, Portugal and [2] Faculty of Medicine, University of Porto, Portugal .

Body: Background: Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disorder that causes severe respiratory dysfunction which is the major cause of death. Early management of respiratory symptoms may improve outcomes and survival. Aim: Describe survival of patients with ALS and respiratory dysfunction and identify predictive factors of survival. Methods: Retrospective analysis of patients with ALS evaluated in an outpatient setting. Ventilatory support data was screened. Kaplan-Meier survival analysis was performed and predictive factors were evaluated by Cox multivariate regression. Results: 60 patients (25 females) with a median age of 64.5 years (range 34-80) were analyzed. At presentation, 33 patients (55%) had slow bulbar-onset and 27 (45%) rapid bulbar-onset. Non-invasive ventilation (NIV) was initiated in 52 patients (86.7%), with a mean vital capacity of 1698.0±768.0 L and 22.0±40.8 months after diagnosis. Mean duration of NIV was 19.6±23.7 months. Mechanical assisted cough was used in 23 patients (38.3%). Gastrostomy was performed in 21 patients (17 rapidly bulbar) and tracheostomy in 10 (9 rapidly bulbar) after a mean time of 13.6 ±17.0 months under NIV. The 5-year survival was 48%. The median overall survival and survival after respiratory muscle aids initiation was significantly higher in slowly bulbar patients compared with rapidly bulbar ($p=0.03$ and $p<0.01$, respectively). In multivariate analysis, predictive factors of survival were younger age, slow bulbar-onset, and early NIV initiation. Conclusion: Survival may be prolonged in patients with ALS and respiratory dysfunction with early NIV initiation. Age and bulbar onset have significant negative impact on survival.