Title: ALS and NIV: A four-year experience in the “NEMO” centre in Italy. A retrospective study

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Body: INTRODUCTION In literature, bulbar ALS patients (pts) have always been related to worse NIV tolerance and survival. There is no clear agreement on NIV parameters to be used. AIM To describe the NIV path in ALS pts attending in the “Nemo” centre, to compare survival differences between onsets, to outline the relationship between respiratory, disability (ALS FRS-R) and NIV parameters. METHOD Our database includes 535 ALS pts, 147 attained our centre between 2008 and 2011 and had their respiratory function been tested from baseline to NIV adaptation and use, until exitus or tracheostomy. From this group, we enrolled 78 pts in a randomized way. Parametric tests were used to compare groups. RESULTS Bulbar pts have the higher diaphragmatic impairment and the worst orthostatic/clinostatic FVC (p=0.038), nevertheless with EPAP titration higher than 4 cmH2O (5.73 ± 1.53) and a proper care of bulbar impairment they can have good tolerance and survive as long as general population. In these pts clinostatic FVC relates directly to ALS FRS-R bulbar and respiratory scores (p<0.01).

CONCLUSION EPAP set higher than 4 cm H2O to reduce ODI, normalize VT and increase SpO2 in NIV permits better tolerance and adherence in all pts. This allows bulbar pts to have a duration of illness from NIV adaptation comparable to other onsets. ALSFRS-R is an accurate measure of respiratory impairment.