**Table. 4 Summary of studies on the use of non-invasive ventilation (NIV) in ALS patients**

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| --- | --- | --- | --- |
| Study | Patients | Results | |
| Mustfa et al. [85] | NIV = **26**  non-NIV = **13**  Control1 = **15**  1 without respiratory muscle weakness | *Survival*  *Quality of life* | **Prolonged** in **NIV** (**NIV** *vs.* **non-NIV**)  ■ median survival time [days]: 298 [95% CI; 192-404] *vs.* 18 [95% CI; 11-25) (p < 0.001)  **No difference** (**NIV** *vs.* **Controls**)  ■ median survival time [days]: 298 [95% CI; 192-404] *vs.* 370 [95% CI; 278-462) (p > 0.05)  **Improved** in **NIV** (**baseline** *vs.* **1-month**) as assessed with **MQOL, SAQLI**, **CRDQ** (fatigue, Emotional, Mastery), **SF-36** (Energy; Vitality) (p < 0.01) |
| Lyall et al. [135] | NIV = **16**  non-NIV = **11** | *Quality of life* | **Improved** (in patients with sleep disorder breathing) |
| Bourke et al. [136] | NIV = **15**/**10** (accepted/continued) | *Quality of life* | **Improved** as assessed with **SF-36 (Mental Component Summary); CRDQ (all domains); SAQLI; ESS.** |
| Pinto et al. [81] | NIV = **10**  non-NIV = **10** | *Survival* | **Improved** in **NIV** (**NIV** *vs.* **non-NIV**)  ■ median survival: [no exact data given] (p < 0.004) |
| Lechtzin et al. [83] | Early NIV1 = **25**  Standard NIV = **67**  1 FVC ≥ 65% predicted | *Survival* | **Prolonged** in **early NIV** (**early NIV** *vs.* **standard NIV**)  ■ median free-tracheostomy survival from the diagnosis [years]: 1.8 vs. 2.7 (p = 0.045) |
| Zamietra et al. [62] | NIV = **6**  NIV + PEG= **5** | *Quality of life* | **No difference** (pre-NIV *vs.* post-NIV)  ■ **ALSSQoL-R** [no data given]  **No difference** (pre-NIV+PEG *vs.* post-NIV+PEG)  **■ ALSSQoL-R** [no data given] |
| Bourke et al. [80] | NIV = **22** (11a+11b)  non-NIV = **19** (9a+ 10b)    a good bulbar function  b poor bulbar function | *Survival (individuals with good bulbar function)*  *Survival (individuals with poor bulbar function)*  *Quality of life (individuals with good bulbar function)*  *Quality of life (individuals with poor bulbar function)* | **Prolonged** in NIV (NIV *vs.* non-NIV*)*  ■ median survival [days]: 216 [range, 94-681] *vs.* 11 [range, 1-283] (p = 0.006)  **No difference** (NIV *vs.* non-NIV)  ■ median survival [days]: 222 [range, 75-1382] *vs.* 261 [range, 6-878] (p = 0.92)  **Improved** in **NIV** (NIV *vs.* non-NIV)  ■ **SAQLI:** all domains  ■ **CRDQ:** all domains  ■ **SF36:** all domains  **No difference** (NIV *vs.* non-NIV)  ■ **SAQLI:** all domains  ■ **CRDQ:** all domains  **■ SF36:** all domains |
| Hein et al. [137] | NIV = **8** | *Quality of life* | *NIV*  **Improved**  ■ **SF-36: mental health**: 55 (SD±13) vs. 64 (SD±17) (p = S) |
| Aboussouan et al. [76] | tolerant of NIV = **23**  intolerant of NIV = **241**  **1** unable to sleep with the device on; interrupted sleep, less than 4 hours per day | *Survival*  *Quality of life (n =* ***8****)* | **Prolonged** in **NIV tolerant** (tolerant of NIV *vs.* intolerant of NIV)  **■** median survival [months]: 20 *vs.* 5 (p = 0.002)  **Improved** in **post-NIV** (pre-NIV *vs.* post-NIV)  **■** CRIQ: mean fatigue score: 11.1 [SD±4.2] *vs.* 14.9 [SD±4.7] (p = 0.03)  **No difference** (pre-NIV *vs.* post-NIV)  ■ CRIQ: mean dyspnea score: 14.7 [SD±3.8] *vs.* 12.6 [SD±3.8] (p = 0.11)  ■ CRIQ: mean dyspnea score: 30.9 [SD±5.5] *vs.* 34.1 [SD±6.3] (p = 0.16)  ■ CRIQ: mean dyspnea score: 16.8 [SD±4.9] *vs.* 19.2 [SD±2.6] (p = 0.07) |
| Burkhardt et al. [82] | NIV = **38**  non-NIV = **33** | *Survival* | **Prolonged** in **NIV** (NIV *vs.* non-NIV)  **■** median survival [months]: 47.7 *vs.* 36.5 (p = 0.01) |
| Kleopa et al. [68] | NIV (>4 hours per day) = **38**  NIV (<4 hours per day) = **32**  non-NIV (refused) = **52** | *Survival*  *Lung function* | **Prolonged** in **NIV (>4 hours per day)** (NIV (>4 hours per day) *vs.*NIV (<4 hours per day)  **■** survival from BIPAP initiation [months]: 14.2 [SD±13.0] *vs.* 7.0 [SD±6.7] (p = 0.002)  **Prolonged** in **NIV (>4 hours per day)** (NIV (>4 hours per day) *vs.*non**-**NIV)  ■ survival from BIPAP initiation [month]: 14.2 [SD±13.0] *vs.* 4.6 [SD±12.7] (p = 0.001)  **Slower** **decline** of VC in **NIV (>4 hours per day)** (NIV (>4 hours per day) *vs.*NIV (<4 hours per day))  ■ slope of %FVC (change/months): 3.5 [SD±5.3] *vs.* 5.9% [SD±4.8] (p = 0.02)  **Slower** **decline** of VC in **NIV (>4 hours per day)** (NIV (>4 hours per day) *vs.*non**-**NIV))  **■** slope of %FVC (change/months): 3.5 [SD±5.3] *vs.* 8.3% [SD±5.0] (p = 0.001) |
| Newsom-Davis et al. [87] | NIV = **9**  Control group1 = **10**  1 ALS patients without symptoms of breathing insufficiency | *Quality of life* | **Improvement** in **NIV at 6th week** (NIV at baseline *vs.* NIV at 6th week)  ■ List learning: 85.58 [SD±9.07] *vs.* 100.13 [SD±8.88] (p = 0.017)  ■ KOLT (raw score): 37.14 [SD±5.84] *vs.* 43.00 [SD+3.37] (p = 0.04)  ■ ESS score: 9.14 [SD±5.27] *vs.* 4.57 [SD±3.82] (p = 0.018)    **No change** in **NIV at 6th week** (NIV at baseline *vs.* NIV at 6th week)  **■** HAD: Anxiety score: 2.80 [SD±3.03] *vs.* 3.40 [SD±1.82] (p = 0.581)  ■ HAD: Depression score: 1.33 [SD±2.31] *vs.* 1.67 [SD±2.89] (p = 0.317) |
| Jackson et al. [71] | Early NIV1 = **6**  1FVC >70% at NIV initiation | *Quality of life* | **Improvement** in **post-NIV** (pre-NIV *vs.* post-NIV) (at 3 month after NIV administration)  ■ SF-36: ‘vitality’ subscale: 10.7 *vs.* 13.0 (p = 0.071)  ■ Pulmonary symptom scale: 72.7 vs. 80.8 (p = 0.04) |

**NIV** = patients who had indication for NIV and decided for; **non-NIV** = patients who had indication for NIV and refused to; **Control group** = patients with no indication for respiratory support; **CRDQ** = Chronic Respiratory Disease Questionnaire; **CRIQ** = Chronic Respiratory Index Questionnaire; **ESS** = Epworth Sleepiness Scale; **HAD** = Hospital Anxiety and Depression Scale; **KOLT** = Kendrick object learning test; **MQOL** = McGill Quality of Life Questionnaire; **SAQLI** = Sleep Apnea Quality of Life Index; **SF-36** = 36-Item Short Form Survey.