Table 1: Description of notification system of the bespoke questions

Table 2: Baseline characteristics of participants (n=13)

Table 3: Median values for each variable over time

Table 4: Results for Wilcoxon tests

Table 5: Intervention category during 24-week trial

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| Table 1: Description of notification system of the bespoke questions | | |
| Notification types | **Definitions** | **Interventions** |
| High priority notifications | Generated for symptoms indicative of hypoventilation, NIV related issues, clearing chest issue, acute respiratory events, swallowing issues, and unmet care needs. | Assessments by clinical team, triggering follow-up intervention as required. |
| Low priority notifications | Generated for symptoms indicative of general deterioration that are not acute. | Further monitoring. |

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| Table 2: Baseline characteristics of participants (n=13) | | |
| *Demographics* |  |  |
| Median age (range) | 66yrs (49yrs-73yrs) |  |
| Male | 8 (61.5%) |  |
| Illness onset |  |  |
| Limb | 11 |  |
| Bulbar | 1 |  |
| Respiratory | 1 |  |
| *Illness characteristics* | *Median (IQR)* | *Range* |
| Illness duration | 14m (39) | (2m–13y7m) |
| NIV duration | 8m (23) | (0–3y) |
| *Baseline function* | *Mean (SD)* |  |
| Respiratory function |  |  |
| FVC % | 55.27 (20.43) |  |
| FEV1/FVC | 82.83 (12.15) |  |
| SNIP% | 45.52 (37.41) |  |

FVC: forced vital capacity; FEV1: forced expiratory volume in one second; SNIP: sniff nasal inspiratory pressure

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| Table 3: Median values for each variable over time | | | |
|  | **Baseline**  **Median (IQR)**  (n=13) | **12 weeks**  **Median (IQR)**  (n=11) | **24 weeks**  **Median (IQR)**  (n=10) |
| ***ALSFRS-R*** |  |  |  |
| Total | 22 (14) | 22 (15) | 20 (21) †‡ |
| Bulbar sub-score | 9 (5.5) | 9 (8.5) | 7 (7.5) † |
| Motor sub-score | 9 (13.5) | 8 (9.5) | 8 (12.5) |
| Respiratory sub-score | 6 (3) | 4 (4) | 4.5 (4.5) |
| ***SpO2*** |  |  |  |
| Average SpO2 levels | 92.5 (5) | 93 (3) | 91 (3.5) |
| Minutes spent <90% | 2.39 (32.99) | 1.33 (12.84) | 12.32 (31.06) |
| Rate of 4% dip | 1.25 (1.44) | 1.05 (3.41) | 1.17 (1.49) |
| ***NIV Use*** |  |  |  |
| Usage time(hrs) | 8 (4) | 11.5 (7) †‡ | 10.5 (7) †‡ |
| ***PVI\**** | **Week 1**  **Median (IQR)** | **Week 12**  **Median (IQR)** | **Week 22**  **Median (IQR)** |
| IPAP | 16.8 (7.7) | 19.17 (10.27) | 21.94 (10.33) †‡ |
| EPAP | 3.97 (0.04) | 3.98 (1.03) | 3.98 (1.02) |
| BPM | 13.63 (3.73) | 14.15 (2.04) | 13.1 (2.81) |
| PTB | 35.92 (63.24) | 47.23 (66.1) | 32.97 (48.53) |
| PF | 22.36 (12.83) | 28.58 (11.29) | 29.71 (10.74) |
| Leak | 38.74 (25.22) | 48.56 (31.95) | 37.05 (38.52) |
| TV | 397.01 (178.74) | 461.59 (257.49) | 528.18 (267.56) |
| TT | 30.7 (6.13) | 32.12 (4.78) | 31.38 (3.83) |
| MV | 5.26 (3.14) | 6.7 (3.21) | 6.62 (4.52) |
| PVI: patient-ventilator interaction; ALSFRS-R: revised Amyotrophic lateral sclerosis functional rating scale; PVI: patient-ventilator interaction; IPAP: Inspiratory positive airway pressure; EPAP: Expiratory positive airway pressure: BPM: Breaths per minute; PTB: Patient triggered breaths; PF: Peak flow; TV: Tidal volume: TT: Ti Titot; MV: Minute ventilation.  \*Data for 24 weeks were obtained from Week 22 due to a low availability of PVI data for Week 23/24.  † Significant changes from the baseline. ‡ Significant changes after Bonferroni correction. | | | |

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| Table 4: Results for Wilcoxon tests | | | |
|  | ***z*-score** | ***r*** | ***p* value** |
| **ALSFRS-R (Total)** |  |  |  |
| Baseline / Week 24 | -2.53 | -.57 | .008 |
| **NIV usage** |  |  |  |
| Baseline / Week 12 | -2.35 | -.48 | .016 |
| Baseline / Week 24 | -2.71 | -.57 | .004 |
| **IPAP** |  |  |  |
| Baseline / Week 22 | -2.52 | -.58 | .008 |

ALSFRS-R: revised Amyotrophic lateral sclerosis functional rating scale; IPAP: Inspiratory positive airway pressure;

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| Table 5: Intervention category during 24-week trial | | |
| **Intervention category** | **Intervention details** | **Number of interventions** (number of participants) |
| Arranging a new appointment  (n=12) | Outpatient appointment | 2 (2) |
| Home visit | 7 (7) |
| Elective admission | 3 (3) |
| Treatment adjustment  (n=16) | NIV adjustment (IPAP increase) | 11 (8) |
| Cough assistor adjustment | 5 (3) |
| Equipment provision  (n=19) | Accessories e.g. masks | 13 (9) |
| Second NIV or exchange NIV | 4 (3) |
| New cough assistor | 1 (1) |
| Humidifier | 1 (1) |
| Referral  (n=14) | Advice from another professionals | 13 (7) |
| GP review advised | 1 (1) |