SUPPLEMENTAL MATERIAL

Supplementary Table 1. Respiratory symptom management characteristics

	US response, *n* (%)	European response, *n* (%)
Pulmonologists only	18 (25.4)	13 (27.1)
Pulmonologists and neurologists	18 (25.4)	16 (33.3)
Neurologists only	15 (21.1)	11 (22.9)
Neurologists and other medical specialists	9 (12.7)	0
Neurologists, pulmonologists, and other medical specialists	8 (11.3)	5 (10.4)
Other medical specialists only	3 (4.2)	2 (4.2)
Other medical specialists and pulmonologists	0	1 (2.1)
Total respondents	71	48

Supplementary Table 2. Always/nearly always responses to the question, "Which of the following are included as part of the evaluation of the respiratory status of your patients with ALS at the time that initiation of NIV is being considered?"

	US response, *n* (%)	European response, *n* (%)
Upright FVC	60 (95.2)	35 (81.4)
Overnight pulse oximetry	5 (7.9)	30 (69.8)
ABGs	2 (3.2)	27 (62.8)
Pulse oximetry	27 (42.9)	21 (48.8)
Supine FVC	20 (31.7)	21 (48.8)
Upright MIP	44 (69.8)	19 (44.2)
SNIP	6 (9.5)	18 (41.9)
Upright SVC	14 (22.2)	17 (39.5)
MEP	24 (38.1)	16 (37.2)
Supine SVC	2 (3.2)	11 (25.6)
Overnight ABGs	0	9 (20.9)
Supine MIP	5 (7.9)	9 (20.9)
Formal sleep study	2 (3.2)	8 (18.6)
Overnight transcutaneous CO_2	0	8 (18.6)
Transcutaneous CO_2	1 (1.6)	4 (9.3)
End tidal CO_2	5 (7.9)	3 (7.0)
Neurophysiological assessment	0	3 (7.0)
Ultrasound assessment of the diaphragm	0	0

ABGs = arterial blood gases; FVC = forced vital capacity; MEP = maximum expiratory pressure; MIP = maximum inspiratory pressure; SNIP = sniff nasal inspiratory pressure; SVC = slow vital capacity.

Supplementary Table 3. The most common subsequent sequence of events

	US response, *n* (%)	European response, *n* (%)
Patient is seen by a pulmonologist/other specialist in the ALS clinic the same day	18 (31.6)	19 (48.7)
Patient is referred to a pulmonologist/other specialist (outside of the ALS clinic) for initiation	15 (26.3)	16 (41.0)
Patient has a trial and is provided instructions on NIV use in the ALS clinic the same day the recommendation is made	5 (8.8)	11 (28.2)
A referral is placed to a home agency and the trial/instructions take place in the patient's home by a respiratory therapist	39 (68.4)	5 (12.8)
Patient is admitted to the hospital for the trial/instructions	0	16 (41.0)
Patient is referred for a sleep study and NIV is titrated in the sleep lab	6 (10.5)	7 (17.9)
Patient is commenced on NIV at home by a specialist outreach nurse	2 (3.5)	5 (12.8)
Total respondents	57	39

ALS = amyotrophic lateral sclerosis; NIV = noninvasive ventilation.

Supplementary Table 4. Preferred type of NIV equipment

	US response, *n* (%)	European response, *n* (%)
Bi-level positive airway pressure with fixed inspiratory and expiratory settings (ex: BiPAP)	8 (14.0)	6 (15.4)
Bi-level positive airway pressure with average volume assured pressure support (ex: BiPAP-AVAPS)	9 (15.8)	3 (7.7)
Ventilators used noninvasively that allow for either pressure or volume-assisted breaths and can be used for mouthpiece ventilation (ex: Trilogy, LTV)	25 (43.9)	5 (12.8)
No preference	4 (7.0)	0
A pulmonologist decides what type of equipment to use	11 (19.3)	25 (64.1)
Total respondents	57	39

NIV = noninvasive ventilation.

Physician Practice Survey on Use of Noninvasive Ventilation in Amyotrophic Lateral Sclerosis

Dear Participants,

This brief (15-20 minutes) survey is being conducted to identify the state-of-the-art practices for use of noninvasive ventilation (NIV) and identify areas where further study may be required. We plan to report our findings at the ALS/MND meeting later this year.
For the purposes of this survey, consider NIV being used for any form of assisted noninvasive ventilation, including instances in which a ventilator capable of providing invasive ventilation is being used non-invasively.
Please only respond to this survey once.

Thank you!

Physician Practice Survey on Use of Noninvasive Ventilation in Amyotrophic Lateral Sclerosis

1. Please specify country of practice:

Country

2. What is your role in the clinic?

Other (please specify)

3. How many individual patients with ALS do you treat in a typical year?

- ◯ 0-25
- ◯ 25-50
- ◯ 51-75
- ◯ 76-100
- ◯ 101-200
- ◯ >200

4. Who manages respiratory symptoms in your clinic?
(*check all that apply*)

- ☐ Neurologist
- ☐ Pulmonologist
- ☐ Other medical specialist

Physician Practice Survey on Use of Noninvasive Ventilation in Amyotrophic Lateral Sclerosis

Even if you do not manage respiratory symptoms yourself, please complete the following questions as best as you can; we want to understand your perception of the state-of-the-art for NIV.

5. Which of the following are included as part of the evaluation of the respiratory status of your patients with ALS **at their initial visit?**

	Always or nearly always	Sometimes (depends upon circumstance/symptoms)	Rarely/Never
Supine FVC	○	○	○
Upright FVC	○	○	○
Supine SVC	○	○	○
Upright SVC	○	○	○
SNIP	○	○	○
Upright MIP	○	○	○
Supine MIP	○	○	○
MEP	○	○	○
End tidal CO2	○	○	○
Transcutaneous CO2 (single value in clinic)	○	○	○
Overnight transcutaneous CO2	○	○	○
Arterial Blood Gases (ABGs)	○	○	○
Overnight ABGs	○	○	○
Pulse oximetry (single value in clinic)	○	○	○
Overnight pulse oximetry	○	○	○
Formal sleep study	○	○	○
Neurophysiological assessment (phrenic nerve conduction study)	○	○	○
Ultrasound assessment of the diaphragm	○	○	○

6. Which of the following are included as part of the evaluation of the respiratory status of your patients with ALS **at their routine follow-up visit(s)?**

	Always or nearly always	Sometimes (depends upon circumstance/symptoms)	Rarely/Never
Supine FVC	○	○	○
Upright FVC	○	○	○
Supine SVC	○	○	○
Upright SVC	○	○	○
SNIP	○	○	○
Upright MIP	○	○	○
Supine MIP	○	○	○
MEP	○	○	○
End tidal CO2	○	○	○
Transcutaneous CO2 (single value in clinic)	○	○	○
Overnight transcutaneous CO2	○	○	○
ABGs	○	○	○
Overnight ABGs	○	○	○
Pulse oximetry (single value in clinic)	○	○	○
Overnight pulse oximetry	○	○	○
Formal sleep study	○	○	○
Neurophysiological assessment (phrenic nerve conduction study)	○	○	○
Ultrasound assessment of the diaphragm	○	○	○

7. Which of the following are included as part of the evaluation of the respiratory status of your patients with ALS **at the time that initiation of NIV is being considered?**

	Always or nearly always	Sometimes (depends upon circumstance/symptoms)	Rarely/Never
Supine FVC	○	○	○
Upright FVC	○	○	○
Supine SVC	○	○	○
Upright SVC	○	○	○
SNIP	○	○	○
Upright MIP	○	○	○
Supine MIP	○	○	○
MEP	○	○	○
End tidal CO2	○	○	○
Transcutaneous CO2 (single value in clinic)	○	○	○
Overnight transcutaneous CO2	○	○	○
ABGs	○	○	○
Overnight ABGs	○	○	○
Pulse oximetry (single value in clinic)	○	○	○
Overnight pulse oximetry	○	○	○
Formal sleep study	○	○	○
Neurophysiological assessment (phrenic nerve conduction study)	○	○	○
Ultrasound assessment of the diaphragm	○	○	○

* 8. Out of the following 17 parameters, please rank your top 7 in importance when making a decision on prescribing NIV. (1=most important, 7=less important). Each number can be used only once.

	1	2	3	4	5	6	7
Supine FVC	○	○	○	○	○	○	○
Upright FVC	○	○	○	○	○	○	○
Supine SVC	○	○	○	○	○	○	○
Upright SVC	○	○	○	○	○	○	○
Supine MIP	○	○	○	○	○	○	○
Upright MIP	○	○	○	○	○	○	○
Overnight pulse oximetry	○	○	○	○	○	○	○
Formal sleep study	○	○	○	○	○	○	○
ABG's in the clinic	○	○	○	○	○	○	○
Overnight ABG's	○	○	○	○	○	○	○
SNIP	○	○	○	○	○	○	○
Symptoms of orthopnea and or dyspnea	○	○	○	○	○	○	○
Sleep related symptoms (e.g. morning headache, snoring, restless sleep)	○	○	○	○	○	○	○
Neurophysiological assessment (phrenic nerve study)	○	○	○	○	○	○	○
Diaphragm ultrasound assessment	○	○	○	○	○	○	○
End tidal CO2	○	○	○	○	○	○	○
Overnight transcutaneous CO2	○	○	○	○	○	○	○

9. Does your country's insurance regulations/national health care coverage impact the following?

	Yes	No	Varies based upon individual patients' coverage
When you initiate NIV	○	○	○
Type of equipment you prescribe	○	○	○
Coverage of NIV equipment if using it less than 4 hrs/24 hrs	○	○	○

Physician Practice Survey on Use of Noninvasive Ventilation in Amyotrophic Lateral Sclerosis

For the following questions, please answer based upon how you currently prescribe NIV, recognizing that these answers may be influenced by insurance/national health care system or other constraints that may be present in your country.

10. For a patient with no respiratory symptoms, at what upright vital capacity (FVC or SVC) would you initiate non-invasive ventilation?

- ◯ <80% of predicted
- ◯ <70% of predicted
- ◯ <60% of predicted
- ◯ <50% of predicted
- ◯ Would initiate NIV at time of diagnosis independent of VC value and symptoms
- ◯ Would not initiate NIV until patient had respiratory symptoms
- ◯ Do not use upright FVC or SVC to decide when to initiate NIV

11. For a patient with respiratory symptoms, at what upright vital capacity (FVC or SVC) would you initiate non-invasive ventilation?

- ◯ <80% of predicted
- ◯ <70% of predicted
- ◯ <60% of predicted
- ◯ <50% of predicted
- ◯ In the presence of respiratory symptoms, would initiate NIV independent of VC value
- ◯ Do not use upright FVC or SVC to decide when to initiate NIV

12. For a patient with no respiratory symptoms, at what supine vital capacity (FVC or SVC) would you initiate non-invasive ventilation?

- ◯ <80% of predicted
- ◯ <70% of predicted
- ◯ <60% of predicted
- ◯ <50% of predicted
- ◯ Would initiate NIV at time of diagnosis independent of VC value and symptoms
- ◯ Would not initiate NIV until patient had respiratory symptoms
- ◯ Do not use supine FVC or SVC to decide when to initiate NIV

13. For a patient with respiratory symptoms, at what supine vital capacity (FVC or SVC) would you initiate non-invasive ventilation?

- ◯ <80% of predicted
- ◯ <70% of predicted
- ◯ <60% of predicted
- ◯ <50% of predicted
- ◯ Do not check VC supine
- ◯ In the presence of respiratory symptoms, would initiate NIV independent of VC value
- ◯ Do not use supine FVC or SVC to decide when to initiate NIV

14. For a patient with no respiratory symptoms, at what upright MIP would you initiate non-invasive ventilation?

- ≤80 cm
- ≤70 cm
- ≤60 cm
- Would initiate NIV at time of diagnosis independent of MIP value and symptoms
- Would not initiate NIV until patient had respiratory symptoms
- Do not use upright MIP to decide when to initiate

15. For a patient with respiratory symptoms, at what upright MIP would you initiate non-invasive ventilation?

- <80 cm
- <70 cm
- <60 cm
- In the presence of respiratory symptoms, would initiate NIV independent of MIP value
- Do not use upright MIP to decide when to initiate NIV

16. For a patient with no respiratory symptoms, at what supine MIP would you initiate non-invasive ventilation?

- ◯ <80 cm
- ◯ <70 cm
- ◯ <60 cm
- ◯ Would initiate NIV at time of diagnosis independent of MIP value and symptoms
- ◯ Would not initiate NIV until patient had respiratory symptoms
- ◯ Do not use supine MIP to decide when to initiate NIV

17. For a patient with respiratory symptoms, at what supine MIP would you initiate non-invasive ventilation?

- ◯ <80 cm
- ◯ <70 cm
- ◯ <60 cm
- ◯ In the presence of respiratory symptoms, would initiate NIV independent of MIP value
- ◯ Do not use MIP to decide when to initiate NIV

18. For a patient with no respiratory symptoms, at what SNIP would you initiate non-invasive ventilation?

- ◯ <80 cm
- ◯ <70 cm
- ◯ <60 cm
- ◯ Would initiate NIV at time of diagnosis independent of SNIP value and symptoms
- ◯ Would not initiate NIV until patient had respiratory symptoms
- ◯ Do not use SNIP to decide when to initiate NIV

19. For a patient with respiratory symptoms, at what SNIP would you initiate non-invasive ventilation?

- ◯ <80 cm
- ◯ <70 cm
- ◯ <60 cm
- ◯ In the presence of respiratory symptoms, would initiate NIV independent of SNP value
- ◯ Do not use SNIP to decide when to initiate NIV

20. If insurance, or other financial constraints were not present, would you alter the timing of when you prescribe NIV?

- ◯ No
- ◯ I would initiate it at the time of diagnosis, independent of symptoms or respiratory testing results
- ◯ I would initiate it at the time respiratory symptoms develop, independent of respiratory testing results
- ◯ Other (please specify in text box below)

Other

21. When you recommend NIV, what is/are the most common subsequent sequence of events? (*check all that apply*)

- ☐ Patient is seen by a pulmonologist/other specialist in the ALS clinic the same day
- ☐ Patient is referred to a pulmonologist/other specialist (outside of the ALS clinic) for initiation
- ☐ Patient has a trial and is provided instructions on NIV use in the ALS clinic the same day the recommendation is made
- ☐ A referral is placed to a home agency and the trial/instructions take place in the patient's home by a respiratory therapist
- ☐ Patient is admitted to the hospital for the trial/instructions
- ☐ Patient is referred for a sleep study and NIV is titrated in the sleep lab
- ☐ Patient is commenced on NIV at home by a specialist outreach nurse

22. What do you consider the minimum goal in hours used per 24 hours in order for the patient to benefit from NIV use?

- ◯ 1 hour/24 hrs
- ◯ 2 hours/24 hrs
- ◯ 3 hours/24 hrs
- ◯ 4 hours/24 hrs
- ◯ >50% night period
- ◯ >90% night period

23. What is your preferred type of NIV equipment?

- ◯ Bilevel positive airway pressure with fixed inspiratory and expiratory settings (ex: BiPAP)
- ◯ Bilevel positive airway pressure with average volume assured pressure support (ex: BiPAP-AVAPS)
- ◯ Ventilators used non-invasively that allow for either pressure or volume assisted breaths and can be used for mouthpiece ventilation (ex: Trilogy, LTV)
- ◯ No preference
- ◯ A pulmonologist decides what type of equipment to use
  (please specify equipment patients are most often prescribed in text box below)

Other

24. What are the obstacles to successful NIV use?

Please assign the level of importance for each of the following:

	Very Important	Important	Somewhat Important	Not Important
Patient not seeing the need	○	○	○	○
Burden for caregiver	○	○	○	○
Cognitive impairment	○	○	○	○
Insurance/National Health System coverage	○	○	○	○
Patient's claustrophobia	○	○	○	○
Inability to effectively manage oral secretions	○	○	○	○
Uncomfortable interface	○	○	○	○
Bulbar symptoms	○	○	○	○
Patient residence area	○	○	○	○
Patient motivation	○	○	○	○

25. Please rank the following as to their importance in your decision regarding recommending NIV.
Rank (1 = most important; 5 = least important)

	1	2	3	4	5
Published guidelines from the AAN	○	○	○	○	○
Published guidelines from the EFNS	○	○	○	○	○
Personal experience/judgment	○	○	○	○	○
Patient wishes/motivation	○	○	○	○	○
Insurance/National Health System coverage	○	○	○	○	○

Comments (optional)