



# The Borg dyspnoea score: a relevant clinical marker of inspiratory muscle weakness in amyotrophic lateral sclerosis

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**ABSTRACT:** The aim of the study was to determine whether the Borg dyspnoea scale could be a useful and simple marker to predict respiratory muscle weakness in amyotrophic lateral sclerosis (ALS).

From April 1997 to 2001, respiratory function was performed in 72 patients together with the Borg score in both the upright (uBorg) and supine (sBorg) positions.

Mean upright vital capacity (VC) was  $81 \pm 24\%$  predicted, sniff nasal inspiratory pressure (SNIP) was  $55 \pm 26\%$  pred, maximal inspiratory pressure ( $P_{I,max}$ ) was  $57 \pm 26\%$  pred and arterial carbon dioxide tension ( $P_{a,CO_2}$ ) was  $41 \pm 6$  mmHg. The mean Borg scores in the upright and supine positions were  $1.7 \pm 1.5$  and  $2.2 \pm 2$ , respectively. A significant relationship between SNIP and uBorg ( $r=0.4$ ;  $p=0.0007$ ) and SNIP and sBorg ( $r=0.58$ ;  $p<0.0001$ ) was observed. Upright VC,  $\Delta$ VC (measured as the supine fall in VC as a percentage of seated VC),  $P_{I,max}$  and  $P_{a,CO_2}$  were significantly correlated with SNIP. A cut-off value of 3 on the sBorg scale provided the best sensitivity (80%) and specificity (78%) (area under the curve 0.8) to predict a SNIP  $\leq 40$  cmH<sub>2</sub>O, indicating severe inspiratory muscle weakness. Patients with a sBorg score  $\geq 3$  also exhibited significantly lower VC,  $P_{I,max}$  and twitch mouth pressure during cervical magnetic stimulation, and slightly higher  $P_{a,CO_2}$  ( $43.7 \pm 7$  versus  $39.2 \pm 5$  mmHg;  $p=0.05$ ).

The Borg dyspnoea scale is a valuable noninvasive test for the prediction of inspiratory muscle weakness in ALS patients.

**KEYWORDS:** Amyotrophic lateral sclerosis, Borg scale, respiratory muscle strength

Amyotrophic lateral sclerosis (ALS) is a devastating and progressive neurodegenerative disorder of unknown aetiology that affects both the upper and lower motor neuron pathways of the nervous system. Respiratory failure (resulting from progressive respiratory muscle weakness) is the usual cause of death [1, 2]. The degree of respiratory muscle involvement is considered difficult to quantify from respiratory symptoms alone. The vital capacity (VC) is the main parameter used in clinical practice to indirectly determine respiratory muscle involvement, but has the disadvantage of a poor sensitivity. The supine position may aggravate symptoms (particularly dyspnoea) and affect spirometry values such as VC. Supine fall in VC was demonstrated to be a sensitive marker of diaphragmatic weakness [3, 4]. The presence of orthopnea was also found to be a marker of poor prognosis in ALS patients

but its intensity was not quantified. The rate of decline of VC is the standard method used to evaluate the progression of respiratory involvement [5–7]. Both volitional tests (such as maximal inspiratory pressure ( $P_{I,max}$ ), maximal expiratory pressure ( $P_{E,max}$ ) and sniff nasal inspiratory pressure (SNIP)) and patient-independent tests (such as twitch mouth pressure ( $P_{m,tw}$ ) or transdiaphragmatic pressure) can be used to assess respiratory muscle strength (RMS) more specifically [8–13]. Recently, MORGAN *et al.* [14] showed that SNIP is a valuable and simple noninvasive test for the detection of inspiratory muscle weakness with a high prognostic value in ALS [14].

Noninvasive positive pressure ventilation (NIPPV) has been shown to improve survival, slow the rate of decline of RMS and improve quality of life in ALS patients [15–18]. Current ALS practice guidelines suggest the monitoring of upright VC in order to detect declining

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respiratory function [6]. These guidelines concur with those of the American College of Chest Physicians, supporting the initiation of NIPPV at the onset of respiratory symptoms, or once a fall below 50% of predicted upright VC and/or a arterial carbon dioxide tension ( $P_{a,CO_2}$ )  $\geq 45$  mmHg and/or a  $P_{I,max} < 60$  cmH<sub>2</sub>O are documented [19].

Many of the aforementioned specific markers of respiratory muscle function are not readily available at the time of initial assessment of ALS patients by the neurologist. Careful evaluation of respiratory signs and symptoms is recommended in current guidelines on ALS management, as defined by the French consensus on ALS management [20]. Little is known about the clinical significance of available dyspnoea scores in ALS, particularly regarding their ability to predict inspiratory muscle weakness. The aim of this study was therefore to evaluate the performance of the modified Borg scale [21] to detect respiratory muscle weakness in ALS patients.

## METHODS

### Study population

From April 1997 to 2001, 72 patients fulfilling the modified El Escorial Criteria [22] were systematically assessed for respiratory manifestations of ALS according to current guidelines [20]. Patients with significant coexisting cardiac or pulmonary disorders were excluded. All data from an individual patient were collected and no patient had received NIPPV or any other ventilatory support (e.g. continuous positive airway pressure (CPAP) and cough assist) at the time of assessment. This retrospective study was approved by the institutional review board of the French language society for respiratory medicine (Société de Pneumologie de Langue Française, Paris, France) and all patients gave informed consent.

### Clinical assessment

A detailed pulmonary evaluation was conducted in order to document the presence of respiratory symptoms and signs of respiratory dysfunction. Prior to pulmonary function testing, subjects were requested to quantify their sensation of dyspnoea by pointing to a score on a large modified Borg scale category ratio 10 (CR10) [21] in both the upright (uBorg) and then in supine (sBorg) positions. The CR10 scale developed by BORG [21] consists of verbal descriptors adjacent to specific numbers, the spacing of the numbers and corresponding descriptors essentially providing a category scale with ratio properties (appendix). The patients were subdivided into bulbar or limb groups as assessed by the Norris score.

### Pulmonary function tests

Spirometry and respiratory muscle assessment were performed with a Medisoft® Hyp'Air system (Medisoft Co Ltd, Dinant, Belgium). VC was measured in both the sitting and supine positions and was expressed as a percentage of the predicted value [23]. The  $\Delta VC$  was calculated as the percentage fall in VC between the supine and upright position [24]. Static mouth pressure was measured using a flanged mouthpiece.  $P_{I,max}$  sustained for 1 s was determined at functional residual capacity, as previously described [8]. SNIP was measured *via* a catheter through an occluded nostril during a maximal sniff [9]. Inspiratory pressures were expressed as a percentage of predicted values as described by ULDRY and FITTING [10]. Static

mouth pressure in response to cervical magnetic stimulation (mouth twitch pressure ( $P_{m,tw}$ )) was measured using a Magstim 200 (Magstim Co. Ltd, Whitland, UK) fitted with a 90-mm circular coil as previously described [11]. A  $P_{m,tw} \geq 12$  cmH<sub>2</sub>O was considered to exclude significant diaphragmatic weakness, as described previously by HUGHES *et al.* [25]. Arterial blood gas analysis was performed in the morning but not immediately upon awakening.

### Nocturnal oximetry

A Novamatrix® pulse oximeter (Novamatrix Inc. Wellingford, CT, USA) was used to record oxygen saturation during sleep and the device recorded oxygen saturation levels at 8-s intervals. Dedicated software (Novacard®; Novamatrix Inc.) determined mean oxygen saturation and the proportion of night-time spent with arterial oxygen saturation ( $S_{a,O_2}$ )  $< 92\%$  and  $< 90\%$ . A percentage of night-time of  $> 5\%$  with a  $S_{a,O_2} < 90\%$  is considered as a criterion to propose NIPPV, according to the French consensus for ALS care [20].

### Follow-up

After a retrospective chart review, 14 patients with a complete respiratory follow-up (including Borg scores) for  $> 1$  yr (mean  $\pm$  SD follow-up  $18.7 \pm 9$  months) were selected to analyse the respective time courses of SNIP and Borg score.

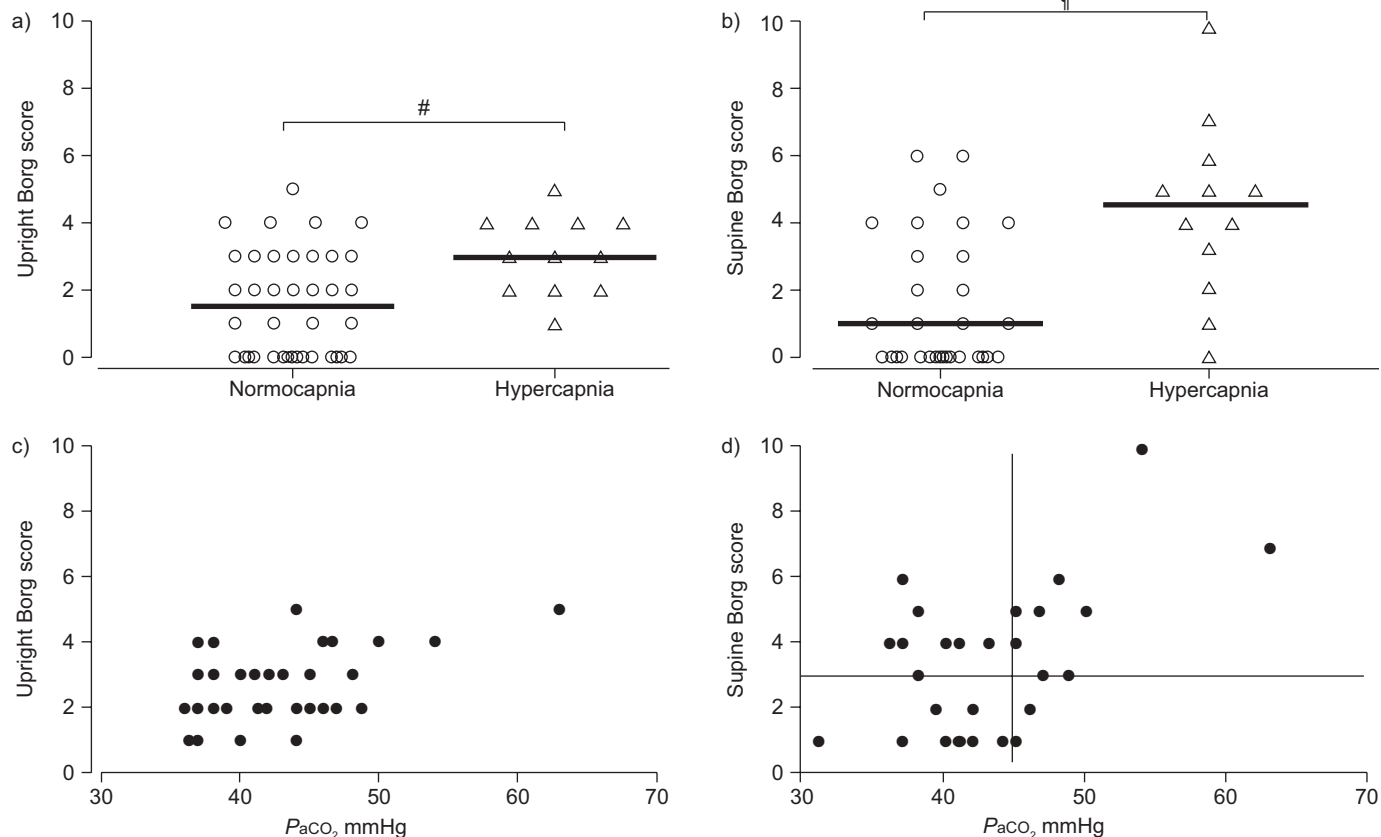
### Statistical analysis

Values are expressed as the mean  $\pm$  SD. Statistical analysis was performed using SPSS software (SPSS, Chicago, IL, USA). Univariate analysis was initially performed to evaluate the

**TABLE 1** Anthropometric and respiratory data for the 72 amyotrophic lateral sclerosis patients

Age yrs	59 $\pm$ 11
Sex males/females n	44/28
BMI kg·m <sup>-2</sup>	25 $\pm$ 4
Exertional dyspnoea	41 (57)
Supine dyspnoea	27 (37)
Respiratory paradox	8 (11)
Accessory muscle recruitment	4 (5.5)
Upright Borg score <sup>#</sup>	1.7 $\pm$ 1.5
Supine Borg score <sup>#</sup>	2.2 $\pm$ 2
$P_{a,CO_2}$ mmHg	41 $\pm$ 6
Upright VC % pred	81 $\pm$ 26
$\Delta VC$ %	-7.7 $\pm$ 9
FEV <sub>1</sub> % pred	80.2 $\pm$ 11
$P_{I,max}$ cmH <sub>2</sub> O	51.8 $\pm$ 27
$P_{I,max}$ % pred	57 $\pm$ 26
SNIP cmH <sub>2</sub> O	51 $\pm$ 24
SNIP % pred	55 $\pm$ 26
$P_{m,tw}$ cmH <sub>2</sub> O	7.8 $\pm$ 4.9

Data are presented as mean  $\pm$  SD or n (%), unless otherwise stated, and were compared with predicted values for pulmonary function and static pressure tests at functional residual capacity. BMI: body mass index;  $P_{a,CO_2}$ : arterial carbon dioxide tension; VC: vital capacity; % pred: % predicted;  $\Delta VC$ : supine fall in VC (percentage of seated VC); FEV<sub>1</sub>: forced expiratory volume in 1 s;  $P_{I,max}$ : maximal static inspiratory pressure; SNIP: sniff nasal inspiratory pressure;  $P_{m,tw}$ : mouth twitch pressure. <sup>#</sup>: out of 10.



**FIGURE 1.** High Borg scores in both positions are associated with hypercapnia. Scatter plots of the Borg score in both a) upright and b) supine positions in amyotrophic lateral sclerosis patients (n=54). The Borg scores were higher in both positions in patients with hypercapnia compared with those with normocapnia. #: p=0.005; †: p=0.003. The lower panels show the relationship between Borg score in both c) upright (R=0.47; p=0.002) and d) supine (R=0.46; p=0.003) positions and hypercapnia. d) Vertical line indicates hypercapnia (45 mmHg); horizontal line indicates supine Borg score cut-off value (3). *P<sub>a</sub>CO<sub>2</sub>*: arterial carbon dioxide tension.

independent contribution of each variable. To limit collinearity in the models, the independent predictor variables were analysed using Pearson correlation coefficients. The predictor variables were included in the models if the correlation coefficient was <0.6. A full model, stepwise, multiple, linear regression was then used to determine the influence of each variable. The level of significance was set at 5%. A receiver operator characteristic (ROC) analysis was performed in order to evaluate the performance of the Borg scale to detect significant inspiratory muscle weakness, defined by a SNIP ≤40 cmH<sub>2</sub>O [14]. Differences between groups were assessed using the Mann–Whitney test. Follow-up SNIP and sBorg scores were compared to baseline values by Wilcoxon signed rank test.

**RESULTS**

**Anthropometric, clinical and respiratory characteristics**

The anthropometric, clinical and respiratory data obtained for our 72 patients are shown in table 1. 16 (22%) patients were bulbar. 41 (57%) patients described exertional dyspnoea. The mean ratings with the Borg scale in both the upright and supine positions were 1.7±1.5 and 2.2±2, respectively. The Borg rating was higher in the supine than in the sitting position in 28 patients. The Borg rating was not significantly different between bulbar and limb subgroups.

**Relationship between Borg score and hypoventilation**

Arterial blood gas measurements were performed in 54 patients. 13 patients (24%) had a *P<sub>a</sub>CO<sub>2</sub>* ≥45 mmHg. Patients with hypercapnia had significantly higher uBorg and sBorg scores (fig. 1a and b). Good correlations between *P<sub>a</sub>CO<sub>2</sub>* and Borg scores were observed in both the upright (r=0.47; p=0.002) and supine positions (r=0.47; p=0.003) (fig. 1c and d). Nocturnal oximetry was obtained for 42 patients. No linear correlation was found between mean or minimal nocturnal *S<sub>a</sub>O<sub>2</sub>* and inspiratory muscle strength or Borg score. Only a weak correlation was found between the percentage of night-time spent with a *S<sub>a</sub>O<sub>2</sub>* <92% and SNIP (table 2).

**Predictive value of Borg score to detect inspiratory muscle weakness**

The results of univariate and multivariate regression analysis are shown in table 2. The Borg score in both the sitting and supine positions showed a significant relationship with SNIP (cmH<sub>2</sub>O). Upright VC % predicted, ΔVC, *P<sub>I,max</sub>* (cmH<sub>2</sub>O) and *P<sub>a</sub>CO<sub>2</sub>* were also significantly correlated with SNIP. sBorg score correlated with SNIP more significantly than *P<sub>m,tw</sub>* but less than *P<sub>I,max</sub>* and VC. The graphical relationships between sBorg score, SNIP and sitting VC are shown in figure 2. In stepwise multiple regression analysis (table 2), *P<sub>I,max</sub>* (cmH<sub>2</sub>O) and sBorg score were both independent predictors of SNIP

**TABLE 2** Results of linear regression to predict sniff nasal inspiratory pressure (in cmH<sub>2</sub>O)

Variables	Coefficient	p-value	R <sup>2</sup>
<i>P</i> <sub>I,max</sub> cmH <sub>2</sub> O	0.73	<0.0001	0.52
<i>P</i> <sub>I,max</sub> %	0.71	<0.0001	0.5
Upright VC %	0.66	<0.0001	0.43
ΔVC %	0.41	0.0016	0.16
<i>P</i> <sub>m,tw</sub> cmH <sub>2</sub> O	0.47	0.002	0.22
Supine Borg score	0.58	<0.0001	0.34
Upright Borg score	0.4	0.0007	0.15
Per cent of night-time with O <sub>2</sub> <92%	0.41	0.016	0.17
Per cent of night-time with O <sub>2</sub> <90%	0.056	0.73	0.003
Mean O <sub>2</sub> saturation %	0.052	0.75	0.003
<i>P</i> <sub>a,CO<sub>2</sub></sub> mmHg	0.26	0.057	0.05
<i>P</i> <sub>a,O<sub>2</sub></sub> mmHg	0.073	0.6	0.005
BMI kg·m <sup>-2</sup>	0.17	0.19	0.03
<i>P</i> <sub>I,max</sub> cmH <sub>2</sub> O, sBorg and ΔVC %		<0.0001	0.67

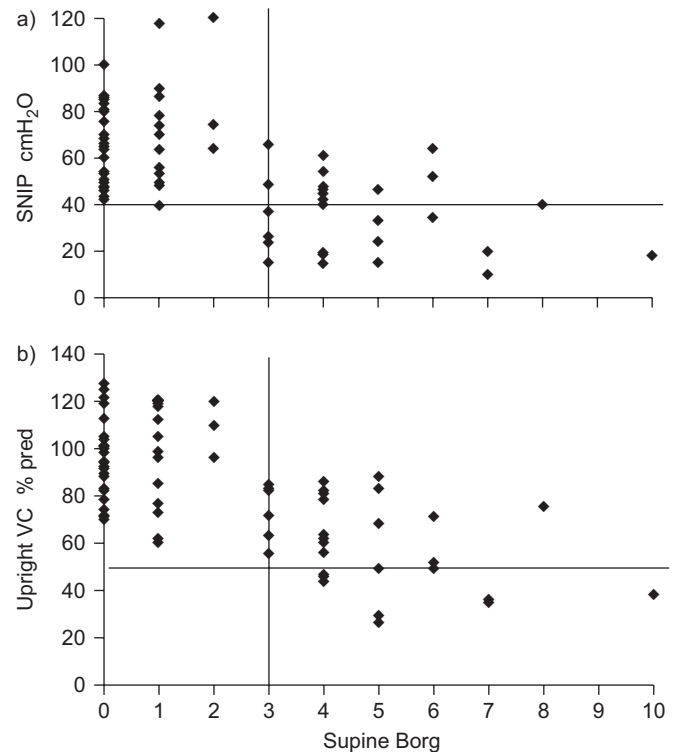
*P*<sub>I,max</sub>: maximal inspiratory pressure; VC: vital capacity; ΔVC: difference from upright VC to supine; *P*<sub>m,tw</sub>: mouth twitch pressure; *P*<sub>a,CO<sub>2</sub></sub>: arterial carbon dioxide tension; *P*<sub>a,O<sub>2</sub></sub>: arterial oxygen tension; BMI: body mass index; sBorg: Borg score in the supine position. p-value reflects F-test for probability of no relationship between predictor variables and sniff nasal inspiratory pressure. R<sup>2</sup> is the coefficient of determination.

(cmH<sub>2</sub>O). The R<sup>2</sup> value in this model of multiple regression was 0.67 (p<0.0001). A model containing *P*<sub>I,max</sub> (cmH<sub>2</sub>O), sBorg score, ΔVC, age, height and weight predicted 71% of the variance of SNIP. The ROC curve graphically presents the performance of Borg scale in the supine position as an indicator of significant inspiratory muscle weakness, defined by a SNIP ≤40 cmH<sub>2</sub>O (fig. 3). The area under the curve was 0.8 and a cut-off value of 3 on the sBorg scale provided the best sensitivity and specificity (80% and 78%, respectively). Using a cut-off value of 3, the Borg scale in the supine position exhibited a good predictive value to detect reduced VC, inspiratory muscle weakness or hypercapnia (table 3). Borg score appeared better than VC to detect a SNIP ≤40 cmH<sub>2</sub>O (sensitivity 62% and specificity 87% for a 70% pred cut-off; and sensitivity 60% and specificity 95% for 50% cut-off). The sBorg is more sensitive than respiratory symptoms on simple enquiry (yes/no). The sensitivity of orthopnea symptoms to detect a reduced SNIP (≤40 cmH<sub>2</sub>O), a VC ≤50% and hypercapnia was 72%, 87.5% and 54%, respectively.

#### Comparison of patients stratified according to sBorg score

28 (39%) patients had a sBorg score ≥3 (group I) whereas 44 (61%) did not describe supine dyspnoea (group II) (table 4). There was no difference in body mass index or proportion of bulbar patients between the two groups. Patients with a high sBorg score were characterised by a lower VC and inspiratory muscle strength, more sleep desaturations (S<sub>a,O<sub>2</sub></sub> <92%), together with a marginally higher daytime *P*<sub>a,CO<sub>2</sub></sub>.

28 patients had an increase in Borg score when changing from sitting to supine posture. All of these patients had significant lower RMS than those with no complaints: mean upright VC 66.7±25 versus 91±19% pred, *P*<sub>I,max</sub> 45±23 versus 66±25%

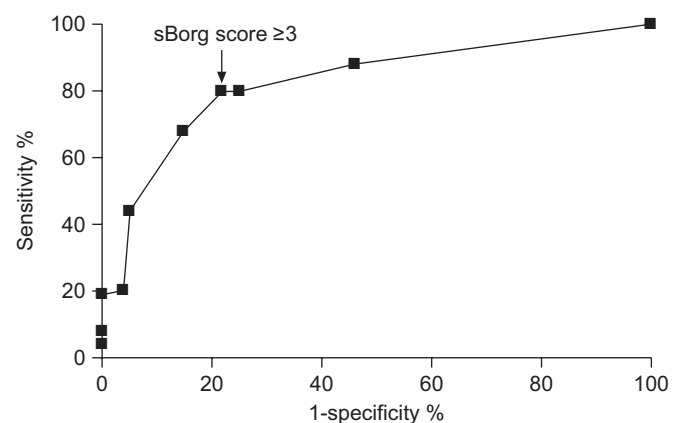


**FIGURE 2.** Relationships between supine Borg and a) sniff nasal inspiratory pressure (SNIP;  $r=-0.58$ ,  $p<0.0001$ ) and b) upright vital capacity (VC;  $r=-0.67$ ,  $p<0.0001$ ). Vertical lines indicate supine Borg score cut-off value (3). Horizontal lines indicate SNIP (40 cmH<sub>2</sub>O) and upright VC (50% pred) cut-off values.

pred, SNIP 41.7±21 versus 63.5±21% pred, *P*<sub>m,tw</sub> 6.2±5 versus 8.7±5 cmH<sub>2</sub>O and ΔVC -6.4±9 versus -10.2±11%.

#### Time course of changes in SNIP and Borg scores

A significant decrease of median (range) SNIP during follow-up (18.7±9 months) was observed: 28.5 (18.5–37.75) versus 58.5 (53–69.5) cmH<sub>2</sub>O (p=0.001). A parallel increase in sBorg score was observed in all patients: 3.5 (3–5) versus 0 (0–0.75) (p=0.0009) (fig. 4).



**FIGURE 3.** A receiver operating characteristic (ROC) curve discriminates sniff nasal inspiratory pressure ≤40 cmH<sub>2</sub>O using supine Borg (sBorg) as an index. The range 0 to 7 was used to determine the appropriate cut-off value. A cut-off value of 3 on the sBorg scale provides the best sensitivity (80%) and specificity (78%).

**TABLE 3** Ability of supine Borg score  $\geq 3$  to predict inspiratory (global or diaphragmatic) weakness and/or hypoventilation

Variables	Subjects n	Sensitivity %	100-specificity %	PPV %	NPV %
SNIP $\leq 40$ cmH <sub>2</sub> O	72	80 (70–89)	22 (12–32)	66 (54–77)	88 (79–96)
VC $\leq 50\%$ pred	72	90 (82–97)	31 (20–42)	32 (20–43)	97 (92–101)
VC $\leq 70\%$ pred	72	80 (70–89)	22 (20–42)	60 (48–72)	97 (92–101)
$P_{m,tw} \leq 11$ cmH <sub>2</sub> O	56	46 (32–60)	7 (0–14)	95 (88–101)	53 (39–67)
$P_{a,CO_2} \geq 45$ mmHg	54	69 (55–82)	27 (18–44)	45 (30–59)	88 (78–97)
$\Delta VC \geq 15\%$	58	66 (52–79)	26 (14–38)	40 (26–53)	89 (80–98)

Data are presented as n (95% confidence interval), unless otherwise stated. PPV: positive predictive value; NPV: negative predictive value; SNIP: sniff nasal inspiratory pressure; VC: vital capacity in sitting position; % pred: % predicted;  $P_{m,tw}$ : mouth twitch pressure;  $P_{a,CO_2}$ : arterial carbon dioxide tension;  $\Delta VC$ : supine fall in VC (percentage of seated VC).

## DISCUSSION

Our study is the first to demonstrate that the Borg dyspnoea score, especially in the supine position, is a noninvasive predictor of respiratory muscular weakness, with a good sensitivity and specificity. These results indicate that the Borg scale should be systematically included in the routine initial assessment and monitoring of ALS patients by the neurologist. This clinical evaluation does not obviate the need for regular pulmonary function tests (PFTs) but may indicate an earlier evaluation. An sBorg value of  $\geq 3$  should prompt PFTs, including arterial blood gases. In contrast to the majority of dyspnoea scales (generally designed for assessment in the context of physical activity [26]), Borg developed a more flexible scale that is particularly well suited to evaluating rapid changes in dyspnoea at a specific time and under a specific set of conditions [21]. Moreover, this scale can easily be scored in patients with limited mobility of the upper extremities and/or who are unable to write. To our knowledge,

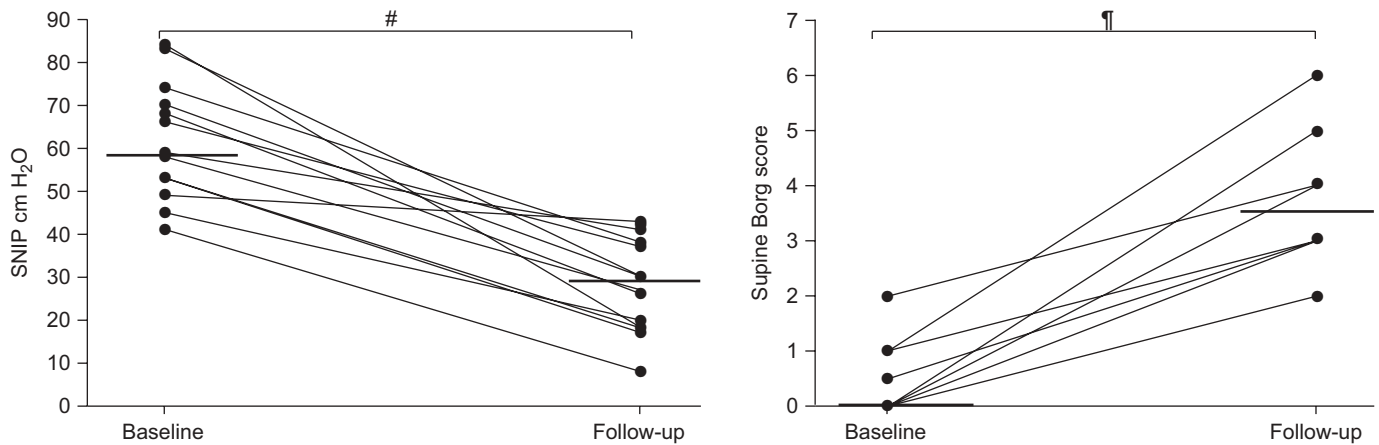
only one study has evaluated a specific dyspnoea scale in ALS [27]. The authors used the baseline dyspnoea index and transition dyspnoea index (BDI-TDI) and found a good correlation with decline in forced VC (FVC) from baseline to follow-up at weeks 4 and 12, better than that of the revised ALS functional rating scale (ALS-FRS-R) and a visual analogue scale. Further studies should compare the predictive value of the Borg score and different dyspnoea scales, such as BDI-TDI and the ALS-FRS-R dyspnoea-orthopnea items. However, the applicability of BDI-TDI scales is limited in ALS patients with severe peripheral muscle weakness, as such nonrespiratory causes of limitation must be quoted Y (non-evaluable) in the questionnaires [26]. Health-related quality of life tools may provide a precise analysis of symptoms and correlate with respiratory muscle performance [28]. This is particularly the case for the dyspnoea domain of the chronic respiratory questionnaire (CRQ). However, these questionnaires are time consuming and are therefore difficult to use

**TABLE 4** Subgroup analysis of amyotrophic lateral sclerosis patients with or without significant supine Borg (sBorg) score

	r	sBorg $\geq 3$ (group I)	sBorg $< 3$ (group II)	p-value <sup>#</sup>
Subjects n		28	44	
pH	-0.13	7.41 $\pm$ 0.02	7.42 $\pm$ 0.03	0.45
$P_{a,O_2}$ mmHg	-0.19	79.9 $\pm$ 14	87.3 $\pm$ 18	0.06
$P_{a,CO_2}$ mmHg	0.33	43.7 $\pm$ 7	39.2 $\pm$ 5	0.05
Mean O <sub>2</sub> saturation %	-0.04	93.8 $\pm$ 2	94.1 $\pm$ 2	0.88
Per cent of night with $S_{a,O_2} < 90\%$	0.15	11 $\pm$ 24	6.2 $\pm$ 16	0.2
Per cent of night with $S_{a,O_2} < 92\%$	0.34	23 $\pm$ 32	6.2 $\pm$ 10	0.01
Upright VC % pred	-0.6	62.5 $\pm$ 18	93.5 $\pm$ 20	<0.0001
$\Delta VC$ %	-0.32	-12.3 $\pm$ 11	-5.3 $\pm$ 8	0.008
$P_{I,max}$ cmH <sub>2</sub> O	-0.48	35.6 $\pm$ 19	62.8 $\pm$ 26	<0.0001
$P_{I,max}$ % pred	-0.47	42 $\pm$ 22	67.6 $\pm$ 24	<0.0001
SNIP cmH <sub>2</sub> O	-0.58	33.8 $\pm$ 15	62.7 $\pm$ 21	<0.0001
SNIP % pred	-0.59	36.6 $\pm$ 16	66.6 $\pm$ 20	<0.0001
$P_{m,tw}$ cmH <sub>2</sub> O	-0.32	5.5 $\pm$ 3.8	9 $\pm$ 5	0.003

Data are presented as mean  $\pm$  SD, unless otherwise stated.  $P_{a,O_2}$ : arterial oxygen tension;  $P_{a,CO_2}$ : arterial carbon dioxide tension;  $S_{a,O_2}$ : arterial oxygen saturation; VC: vital capacity; % pred: % predicted;  $\Delta VC$ : supine fall in VC (percentage of seated VC);  $P_{I,max}$ : maximal inspiratory pressure; SNIP: sniff nasal inspiratory pressure;  $P_{m,tw}$ : mouth twitch pressure. r is the coefficient of correlation between the supine Borg score and the different respiratory parameters (Spearman correlation test). #: Mann-Whitney test for continuous variables.





**FIGURE 4.** Time courses of sniff nasal inspiratory pressure (SNIP) and supine Borg scores from 14 patients. The SNIP values decreased over time whereas supine Borg score increased. #:  $p=0.001$ ; †:  $p=0.0009$ .

on a daily clinical basis. In comparison, the Borg scale may be administered in <1 min.

A limitation of our study is the lack of comparison of the Borg score with ALS-FRS-R [29] to detect inspiratory muscle weakness. The ALS-FRS-R includes three respiratory clinical items (dyspnoea, orthopnea and respiratory insufficiency) and it would be interesting to assess and compare the correlation between RMS and ALS-FRS-R *versus* sBorg score in a complementary prospective study that could also provide an external validation of Borg score in an independent population.

Previous studies have also reported the utility of postural change in spirometric parameters in the assessment of diaphragmatic function in neuromuscular disease [3]. VARRATO *et al.* [4] showed that in symptomatic ALS patients (*i.e.* those with dyspnoea and/or orthopnea), the supine FVC was significantly lower and the erect–supine FVC difference was significantly greater. The low Borg scores in the sitting position demonstrate the poor sensitivity of daytime dyspnoea as a marker of disease severity in ALS, due to its late occurrence. This finding is reinforced by the weak correlation between uBorg score and other RMS parameters. However, when the symptom score and physical examination are completed with the patient in the supine position, they become much better discriminators of RMS, particularly when evaluating the effects of orthostatic changes on dyspnoea intensity.

As expected, the correlation between Borg score and RMS is stronger for SNIP than for other patient-dependent tests. LYALL *et al.* [13] have previously shown that SNIP provides a more accurate reflection of inspiratory muscle strength, regardless of whether the weakness is of central or peripheral origin. Recently, MORGAN *et al.* [14] showed that SNIP is a better noninvasive test than VC for monitoring RMS, as it has a much higher prognostic value. A SNIP <40 cmH<sub>2</sub>O has a sensitivity of 97% and a specificity of 79% to predict death within 6 months. The sBorg score was less closely correlated with non-volitional tests than with volitional ones. This may be explained by reflex glottic closure during magnetic phrenic nerve stimulation, leading to impaired transmission of pleural and tracheal pressure to the mouth. To limit glottic closure, we performed magnetic stimulation during a gentle expiration

from the functional residual capacity, as described previously [30]. Trans-diaphragmatic pressure with magnetic stimulation is the gold standard for the evaluation of diaphragmatic strength, but this test cannot be performed on a routine basis in the follow up of ALS patients.

The relationship between  $P_{a,CO_2}$  and Borg score is not significant. In concordance with LYALL *et al.* [13], we also observed a threshold of RMS below which ALS patients developed hypercapnia. Previous studies have shown a poor correlation between RMS and  $P_{a,CO_2}$ , particularly when respiratory muscle weakness is quite severe or in patients with bulbar involvement [13]. This may be explained, at least partially, by compensatory hyperventilation that occurs early in the disease, when there is only a moderate degree of respiratory muscle weakness. This lack of correlation may also be explained by other factors, such as the timing of arterial blood gas measurement, which was not performed upon arousal in our population. Hence, some patients may present with isolated nocturnal hypercapnia, overlooked by daytime arterial blood gases. Nocturnal recording of transcutaneous CO<sub>2</sub> may prove more sensitive, as hypoventilation first appears during rapid eye movement sleep in neuromuscular disorders [31]. However, this technique has not been studied extensively in ALS patients. It would also be interesting to perform polysomnography in all patients. Indeed, previous studies have demonstrated that ALS patients develop sleep-disordered breathing with obstructive (but more commonly central) apnoeas and concomitant hypercapnia, although the prevalence of these respiratory events remains highly variable among studies [12, 13].

NIPPV prolongs survival and ameliorates quality of life in patients using it for >4 h per day [16–18]. Therefore, early detection of inspiratory weakness with nocturnal desaturations is likely to improve the management and, thereby, the prognosis of ALS patients. A significant sBorg score corresponded to an increased percentage of sleep desaturations ( $S_{a,O_2} < 92\%$ ) in our patients. This desaturation pattern can be considered as significant in comparison with oximetry findings in normal subjects [32]. The indication of NIPPV initiation is well defined by recent guidelines, which recommend that it is

started in the presence of respiratory symptoms with either inspiratory muscle weakness (defined by  $P_{I,max} \leq 60$  cmH<sub>2</sub>O or VC  $\leq 50\%$  pred), daytime hypoventilation ( $P_{a,CO_2} \geq 45$  mmHg) or nocturnal desaturations [18, 19]. sBorg score demonstrated a high sensitivity (90%) for the detection of an upright VC  $< 50\%$  pred.

In conclusion, the Borg dyspnoea scale is a useful clinical tool for the detection of ventilatory failure in ALS patients. It is particularly suitable for the routine clinical evaluation of respiratory status by the neurologist. The finding of a significant Borg score should prompt a rapid evaluation of respiratory status by performing respiratory muscle tests together with a sleep study. Additional studies are required to compare the sBorg score with the ALSFRS-R respiratory items and to evaluate its performance score in other neuromuscular disorders with respiratory involvement.

## APPENDIX

The category ratio scale modified for use in measuring dyspnoea is shown in table 5.

**TABLE 5** The 0 to 10 category ratio scale modified for use in measuring dyspnoea

0	Nothing at all
0.5	Very, very slight
1	Very slight
2	Slight
3	Moderate
4	Somewhat severe
5	Severe (heavy)
6	
7	Very severe
8	
9	
10	Very, very severe (almost max)

This scale incorporates non-linear spacing of verbal descriptions of severity, which correspond to specific numbers for intensities.

## STATEMENT OF INTEREST

None declared.

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