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RESEARCH ARTICLE

The importance of maintaining the same order of performance of lung function and SNIP tests in patients with amyotrophic lateral sclerosis

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Abstract

Objective: Sniff nasal inspiratory pressure (SNIP), a useful tool for the assessment of diaphragm function in patients with Amyotrophic Lateral Sclerosis (ALS), is usually performed together with lung function tests. The aim of this study was to evaluate whether SNIP results are influenced by the order of performance of the tests. *Methods*: 103 consecutive patients (65% males, 80% spinal onset) were recruited. The highest value of up to 10 sniffs, was recorded before (SNIPT0) and after (SNIPT1) the assessment of lung function, peak cough (PCF), and peak expiratory flow (PEF). *Results*: Mean and median values were respectively 31.10 and 26.00 cm H_2O for SNIPT0 and 28.93 and 25.00 cm H_2O for SNIPT1 ($p < 0.001$). The median value of (SNIPT1 − SNIPT0)/SNIPT0% was −7.10. Patients showing post lung function SNIP values above the median were included in Group 1 (51%), the others in group 2 (49%). Group 2 showed more severe baseline ventilatory restriction and reduction in PCF than Group 1. Positive direct relationships were found between SNIPT0 and SNIPT1 (coefficient $\beta = 0.95$, $p < 0.001$), and forced vital capacity and forced expiratory volume at one second. *Conclusions*: 50/103(49%) patients with ALS show a significant reduction in SNIP when assessed shortly after the performance of lung function tests. These patients suffer from more severe ventilatory restriction than patients not showing the reduction. Our findings suggest standardizing the order of respiratory tests during the follow up in order to avoid to misestimate the real strength of inspiratory muscles.

Keywords: *Amyotrophic lateral sclerosis, respiratory muscle, motoneuron disease, neuromuscular disease, lung function tests*

Introduction

Amyotrophic lateral sclerosis (ALS) is a rare, progressive, and fatal motoneuron disease with 2–4 years average timeframe between symptom onset and death, although 5–10% of patients may survive longer (1). Respiratory muscles become weaker and patients progressively develop nocturnal hypoventilation and hypercapnic respiratory failure (2,3). Death is usually related to complications such as acute respiratory failure and lower respiratory tract infections (4–6). Close clinical and functional follow-up is crucial: guidelines suggest a four-monthly multidisciplinary check with particular care for the respiratory system (1). The recommended respiratory monitoring consists in the assessment of lung and respiratory muscle function, as well as of cough efficiency (7,8).

The assessment of sniff nasal inspiratory pressure (SNIP), a method to measure inspiratory muscle strength, has been proposed as an alternative to or a complement of maximal static inspiratory pressures (9,10). The pressures generated

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during these two maneuvers are the result of different complex interactions between several muscle groups since these efforts produce different mechanisms of activity of inspiratory and expiratory muscles. The SNIP test and the maximal static inspiratory pressure when performed with the Mueller maneuver, should be used primarily to assess inspiratory muscle reserve (11).

In patients with ALS, SNIP value has been considered as an important predictive factor of sleep-disordered breathing and need of tracheotomy, and although SNIP used alone is not considered an adequate indication for NIV, it has shown the steepest decline prior to NIV indication (7,8,11–14). Therefore, appropriate monitoring of the time course of SNIP has clinical relevance: as a result, we must warrant good repeatability of the used tests.

A study in patients with neuromuscular diseases other than ALS showed that the performance and results of SNIP can improve with learning and that 10 or more maneuvers should be performed in these patients to reach the maximal result (15). In clinical practice, SNIP is usually performed in the same session of lung and other respiratory muscle function tests. However, in patients with ALS, the performance of these tests may be challenging, potentially leading to fatigue or even exhaustion, eventually influencing the performance of the next volitional tests such as SNIP. Therefore, we hypothesized that the order of the performance of SNIP prior to (SNIP Time 0, T0) or after lung function tests (SNIP Time 1, T1) would influence its results.

Methods

Study population

This prospective study was conducted from January to June 2017 in the Respiratory and Sleep Disorders Unit, University Hospital, Bari, Italy, in 103 patients with a confirmed diagnosis of ALS according to the revised El Escorial criteria (16,17) as part of their four-monthly respiratory follow up. All patients provided written consent to the collection and the scientific use of data. The study was approved by the local Ethic Committee: (no. 5974, 12 June 2019).

Study protocol

One-hundred and three consecutive patients were included in this study and underwent all tests in the same following order: baseline SNIP (SNIP T0), lung function tests (Spirometry: flow/volume curve), Peak cough flow (PCF), Peak expiratory flow (PEF), post-tests SNIP (SNIP T1).

Measurements

The SNIP test was performed in the sitting position (9,10) by means of a Micro RPM-Respiratory Pressure Meter connected to PUMA software (Carefusion Germany 234 GmbH Customer Service & Support International, Hoechberg, Germany). The test was performed during a quick and maximal inspiratory effort with closed mouth via a probe inserted in one nostril. The other nostril was occluded by either the subject or the operator depending on the motor ability of each subject. In our center, the SNIP closed maneuver was opted for as a more useful test to rule out inspiratory muscle weakness (18). The size of the probe was the closest to the subject's nostril among three options (small, medium, large), and connected to a pressure sensor, data being analyzed by specific software. After each maneuver, a visual feedback with the pressure level achieved was visualized on the computer screen by both the subject and the operator in order to motivate patients to their best effort. Patients were controlled by the same operator who set the time between efforts with clear and loud commands, and continuous encouragement to the greatest effort. Maximal inspiratory maneuvers started at the end of a normal expiration. The highest value from at least 8 to 10 maneuvers was recorded.

Lung function tests were performed according to the ERS/ATS Task Force (19). Dynamic volumes were assessed by means of a spirometer (Master Screen PFT system powered by Sentry Suite Carefusion, 234 GmbH Leibnizstrasse 7 Hoechberg Germany). Predictive values were those of Quanjer et al. (20). A forced vital capacity (FVC) $<80\%$ of predicted was considered as representative of a restrictive ventilatory pulmonary defect in presence of normal Forced expiratory volume in one second (FEV1)/Forced Expiratory Capacity (FVC) ratio (21).

PEF and PCF were performed according to the ERS/ATS guidelines (9,10). The voluntary cough was usually initiated at the end of a normal expiration, and 3 to 5 repeated maneuvers were performed for each subject with encouragement and visual feedback. The best value was then reported.

There was no resting time within the same test session (i.e. between each repeated SNIP maneuver). The few minutes necessary for the organization, elapsed between different tests within each patient's session.

Statistical analysis

Changes in the post to pre lung function SNIP are shown as % of baseline values: (SNIP T1-SNIP T0)/SNIP T0% Median value and interquartile ranges (IQR) of all (SNIPT1 – SNIPT0)/SNIPT0% values were calculated. Patients

Table 1. Demographic, anthropometric clinical and physiological characteristics of all subjects.

Variable	
Age (years)	65.3 ± 10.9
Males (%)	65.6
BMI (Kg/m^2)	25.5 ± 4
Non smokers, *n* (%)	60 (58.2)
Smokers, *n* (%)	14 (13.6)
Former smokers, *n* (%)	26 (25.2)
Disease Length, months [IQR]*	43.0 [67.5]
Bulbar onset (%)	19.4
Spinal onset (%)	80.5
No NIV (%)	73.7
NIV (%)	26.2
FEV_1 (% pred.)	72.3 ± 26.3
FVC (% pred.)	70.7 ± 26.2
FEV_1/FVC (%)	79.1 ± 11.9
No restriction (%)	35.9
Restriction (%)	64.1
PEF (L/min)	278.3 ± 139.9
PCF (L/min)	244.6 ± 125.6
SNIPT0 (cm H_2O)	31.1 ± 17.1
SNIPT1 (cmH_2O)	28.9 ± 16.9
Worseners, *n* (%)	50 (49%)
Stables, *n* (%)	53 (51%)
pH	7.4 ± 0.03
P_{aCO2} (mmHg)	41.9 ± 6.7
P_{aO2} (mmHg)	80.1 ± 11.8
S_{aO2} (%)	95.9 ± 1.7

Values are shown as mean ± SD respectively for normal distributed numeric variables or as median [interquartile range] for non-normal distributed numeric variables (*), and with % for categorical ones. Abbreviations. BMI: Body Mass Index; NIV: noninvasive ventilation; P_{aCO2}: arterial carbon dioxide tension; P_{aO2}: arterial oxygen tension; PCF: peak cough flow; PEF: peak expiratory flow; S_{aO2}: arterial oxygen saturation; SNIP: Sniff nasal inspiratory pressure.

showing an increase in SNIP post lung function and those with values greater than the median values of (SNIPT1 − SNIPT0)/SNIPT0% were included in Group 1. The others showing values lower than the median values of (SNIPT1 − SNIPT0)/SNIPT0% were included in Group 2.

Statistical analyses were performed using the R statistical environment, in detail the packages "stats," "fBasics," "car" (22–24). Each variable was reviewed via the Shapiro–Wilk test and graphical evaluations to confirm normal distribution. As a result, all quantitative variables but the length of disease were considered as normally distributed. Descriptive statistics are presented as mean ± standard deviation (SD) and/or median and interquartile range for normally distributed and non-normally distributed continuous variables respectively, whereas categorical variables are indicated with frequency (%) (Table 1).

Student t-test and Mann-Whitney's U test were performed to assess statistical significance between groups, Comparison among SNIP T0 and SNIPT1 was made via t-test for paired groups. Differences between groups of categorical variables were assessed by a Pearson χ^2 test or Fisher's Exact test according to the relative frequencies. Plots and graphs were realized using the R package "graph." Bland Altman plot was realized with package "BlandAltmanLeh" and "ggplot2" (25,26).

Receiver–operating characteristic (ROC) curves were performed using the R package "pROC" (22). "pROC" was also used to assess the power of the obtained ROC curves, to evaluate values of Area Under Curve (AUC), sensitivity, and specificity and to compare ROC curves. An AUC > 0.9 defined a high diagnostic accuracy, a value between 0.7 and 0.9 defined a moderate level of accuracy, whereas a lower AUC indicates a low diagnostic accuracy.

Results of univariate models with regression analyses are shown as regression coefficients β with their 95% confidence intervals. A p value <0.05 was considered as statistically significant.

Results

The characteristics of patients in the study are shown in Table 1. Mean SNIPT0 was significantly higher than SNIPT1. Demographic, anthropometric clinical and physiological characteristics of patients according to Groups are shown in Table 2.

ROC curves were analyzed looking for a precise cut off, and the AUC Delta SNIP% was found at 0.591 (95% confidence interval: 0.47–0.70). Then, median and quartiles (IQR) assessment has been performed, finding quartiles at −0.65, −7.10, and 16.32. Therefore, the median value of (SNIPT1 − SNIPT0)/SNIPT0% was found at −7.10. As shown in Figures 1 and 2, 53 out of 103 (51%) of patients showed a value post lung function test (SNIPT1 − SNIPT0)/SNIPT0% above than median and allocated in group 1, while 50 out of 103 (49%) of patients showed a post lung function test (SNIPT1 − SNIPT0)/SNIPT0% below than median and allocated in group 2. Dynamic volumes, PCF, and PEF were significantly lower in Group2 (Table 2).

Weak direct relationships were found between SNIP T0 and SNIP T1, and between both FVC and FEV1, and SNIPT0, and SNIPT1 but not (SNIPT1 − SNIPT0)/SNIPT0% (Table 3).

Discussion

This study has shown that 50 out of 103 (49%) patients with ALS (group 2) show a significant reduction in SNIP when assessed shortly after the performance of lung function tests. These patients suffer from more severe ventilatory restriction as indicated by lower dynamic volumes with normal FEV1/FVC ratio, as compared with the other

Table 2. Demographic, anthropometric clinical and physiological characteristics of subjects according to Groups.

	(SNIPT1 − SNIPT0)/SNIPT0%		
Variable	Group 1 ($n = 53$)	Group 2 ($n = 50$)	p Value
Age (years)	65.27 ± 10.22	65.42 ± 11.69	0.94
Male, *n* (%)	37 (37.4)	28 (28.3)	0.04
Female, *n* (%)	13 (12.1)	22 (22.2)	
BMI (Kg/m^2)	26.5 ± 5.3	25.3 ± 3.7	0.65
Non smokers, *n* (%)	26 (26)	34 (34)	0.25
Smokers, *n* (%)	9 (8)	6 (6)	
Former smokers, *n* (%)	18 (15)	10 (10)	
Disease Length, months [IQR]*	43.5 [51.0]	42.0 [71.0]	0.48
Bulbar onset, *n* (%)	11 (10.7)	9 (8.7)	0.51
Spinal onset, *n* (%)	42 (40.8)	41 (39.8)	
No NIV, *n* (%)	41 (39.8)	35 (34.0)	0.50
NIV n (%)	12 (11.7)	15 (14.6)	
FEV_1 (% pred.)	78.4 ± 24.4	66.5 ± 27.1	0.02
FVC (% pred.)	76.2 ± 25.2	65.4 ± 26.4	0.04
FEV_1/FVC (%)	80.1 ± 11.3	78.2 ± 12.6	0.44
No restriction, *n* (%)	23 (22.3)	14 (13.6)	0.10
Restriction, *n* (%)	30 (29.1)	36 (35)	
PEF (L/min)	308.6 ± 127.7	247.9 ± 146.4	0.04
PCF (L/min)	276.1 ± 125.1	211.8 ± 118.9	0.01
pH	7.44 ± 0.02	7.44 ± 0.03	0.91
P_{aCO2} (mmHg)	41.5 ± 5.8	42.4 ± 7.6	0.51
P_{aO2} (mmHg)	82.2 ± 11.1	78.1 ± 12.3	0.09
Hco_3-	28.5 ± 3.3	28.5 ± 3.1	0.96
S_{aO2} (%)	96.2 ± 1.5	95.7 ± 1.9	0.17

Group 1= (SNIPT1 − SNIPT0)/SNIPT0 % above the median. Group2 = (SNIPT1 − SNIPT0)/SNIPT0 % below the median. Values are shown as mean ± SD for normal distributed numeric variables or as median [inter-quartile range] for non-normal distributed numeric variables (*), and with % for categorical ones. p Values are adjusted for multiple comparison via Bonferroni correction for non categorical variables. Abbreviations. BMI: Body Mass Index; NIV: non-invasive ventilation; P_{aCO2}: arterial carbon dioxide tension; P_{aO2}: arterial oxygen tension; PCF: peak cough flow; PEF: peak expiratory flow; S_{aO2}: arterial oxygen saturation SNIP: sniff nasal inspiratory pressure.*

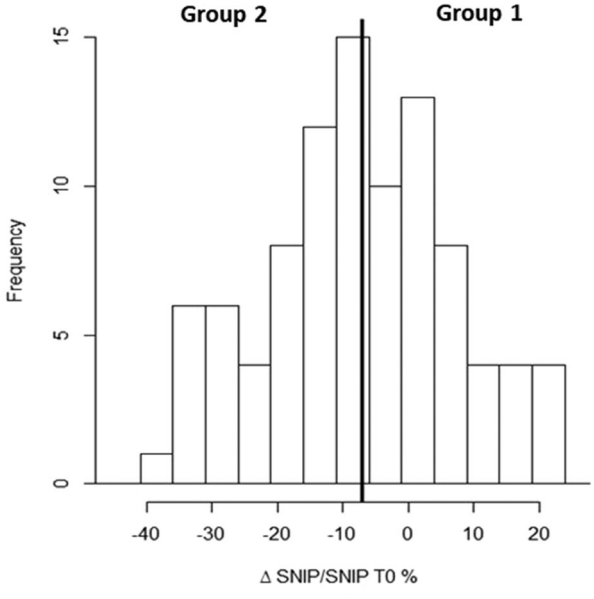

Figure 1. Frequency of representation of (SNIPT1 − SNIPT0)/SNIPT0% among all patients.

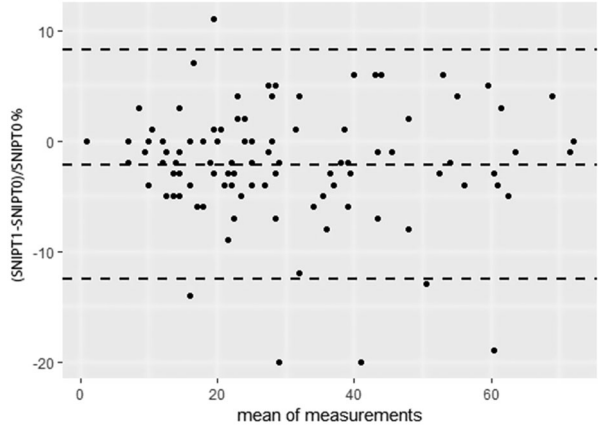

Figure 2. Bland–Altman plot of (SNIPT1 − SNIPT0)/SNIPT0% among all patients.

patients (group 1). Therefore the same order of performance of respiratory muscle and lung function tests should be maintained during the first evaluation and the follow-up visits. To the best of our knowledge, this is the first study to explore this issue.

Sniff nasal inspiratory pressure is a reproducible test over time in healthy people (27). However, reliability has not been assessed in ALS patients. As compared with FVC, SNIP provides more accurate prognostic information on mortality and indication of non-invasive ventilation (NIV) in patients with ALS (7,8). Despite its usefulness, this test is not widely performed on a routine basis

Table 3. Correlation between SNIP results and dynamic volumes among all patients.

	FVC (coefficient β)	*P*	FEV1 (coefficient β)	*p*
SNIPT0	0.46	<0.001	0.42	<0.001.
SNIPT1	0.45	<0.001	0.42	<0.001
(SNIPT1 − SNIPT0)/SNIPT0%	−0.03	0.67	−0.05	0.428

(8,28). A nationwide survey of current Dutch practice reported that SNIP was performed only by 13% of teams assessed, as compared to 72% for FVC and 31–28% of maximal inspiratory and expiratory pressure (29). SNIP was reported as more sensitive than maximal static respiratory pressures in evaluating the diaphragm strength (11,30,31).

In order to exclude any difference in learning effect, in our study the highest value of at least 8–10 maneuvers were recorded each time, however, in almost half of patients, there was a significant drop in SNIP performed after lung function tests. This result suggests performing the lung function and respiratory muscle tests in the same order in each visit along with a follow up to avoid overestimating a reduction in SNIP value.

Rather interestingly FEV1, FVC as well as PCF were significantly lower in Group 2 (Table 2). This observation may indicate that ALS patients with more advanced disease, at least in terms of more severe ventilator restriction require greater attention to the order of assessments during follow up.

The results of this study may have clinical implications. Respiratory muscle weakness often starts as nocturnal hypoventilation during sleep, and symptoms can be treated with NIV, which improves survival and quality of life in selected patients (32–34). Initiation of NIV is usually based on daytime tests of respiratory muscle and lung function, FVC being the most frequently used test in these patients (35,36). Peak cough flow estimates cough efficacy and airway clearance and may help decisions regarding cough augmentation via the use of Cough Assist dedicated machines but its role in predicting the need for NIV is not yet fully clarified (37). The use of SNIP decline has also been suggested as part of routine care in patients with ALS as a sensitive tool to evaluate the diaphragm strength and to avoid non-timely initiation of NIV (7,8,12–14,38,39). As a consequence, we need to ensure that any observed decline over time in SNIP value is actually due to a reduction in respiratory muscle force and progression of the disease and not to differences in the timing of performing the maneuver. Furthermore, a recent study evaluated the SNIP test in ALS and Primary Lateral Sclerosis (PLS) patients via the occluded vs non-occluded measurement mode over a period of 6 years. Results showed that there was an average decline of 0 to10 cm H20 over 3 months period, but some increases were also observed mainly in spinal onset ALS patients (40). This is in line with our results. The authors speculated that could have been related to temporary nasal congestion. However, it could have also been related to the timing in the SNIP performance with respect to other respiratory tests. As a consequence, patients could have resulted in less or more tired by the performance of the spirometry, resulting in higher or lower SNIP values. Hence, our results indicate that the same order of test performance should be applied to all ALS patients, and it is particularly important in patients with more advanced disease. Therefore, the standardization of maneuver is warranted, including the order of performance of tests in different sessions during the follow up over time.

Conclusions

This study has shown that 50 out of 103 (49%) patients with ALS show a significant reduction in SNIP when assessed shortly after the performance of lung function tests. These patients suffer from more severe ventilatory restriction as compared with patients who do not show such reduction. Our findings suggest standardizing the order of respiratory tests during the follow up of these patients in order to avoid misestimating the real strength of inspiratory muscle function.

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Declaration of Interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of this article.

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