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Long-term home noninvasive ventilation (LTHNIV) in restrictive thoracic diseases: The Italian snapshot

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Ethics approval and consent to participate: ERS scientific committee validated the survey before its submission to ERS Assembly 2.02 (NIV dedicated group); participants were waived to submit any consent to participate in the survey, since the survey was anonymous and sensitive data were not collected. All methods were carried out in accordance with relevant guidelines and regulations.

Consent for publication: not required applicable

Authors' contributions: PP, CC, conceptualization; LP, methodology, formal analysis, visualization; AN, software, data curation; RS, AC, GEC, validation; PP, investigation, original manuscript drafting; CC, resources; PP, CC, AC, writing-review and editing; AC, supervision; RS, GEC, project administration. All the authors read and approved the final version of the manuscript and agreed to be accountable for all aspects of the work.

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Abstract

Long-term home noninvasive ventilation (LTHNIV) in restrictive thoracic diseases was explored via the recently published international REINVENT ERS survey. The Italian subset of respondents (ITA-r), the highest above all participating nations, was analyzed and compared to non-Italian respondents (NO-ITA-r). The ITA-r represented 20% of the total answers examined. Ninety-four percent were physicians, whose half worked in a respiratory ICU (RICU). ITA-r mainly worked in community hospitals *vs* NO-ITA-r who are largely affiliated with university hospitals ($p<0.0001$). Amyotrophic lateral sclerosis (ALS) was considered the most common medical condition leading to NIV indication by both ITA-r and NO-ITA-r (93% *vs* 78%, $p>0.5$). A greater proportion of ITA-r considered MIP/MEP the most important test for NIV initiation as compared to NO-IRA-r ($p<0.05$). There was no significant difference for both ITA-r and NO-ITA-r as regards the other questions. This study illustrates Italian LTHNIV practices in patients with NMD and it shows some important differences with the other countries' practices but agreement in terms of goals to achieve, reasons to initiate NIV, and practices among the two communities.

Key words: Noninvasive ventilation; neuro-muscular disorders; amyotrophic lateral sclerosis; ALS; REINVENT.

Introduction

In the last four decades noninvasive ventilation (NIV) has become the reference treatment for the chronic respiratory failure of both obstructive and restrictive thoracic disorders (RTD). This allowed a drastic decrease in home invasive mechanical ventilation while improving survival,

quality of sleep and quality of life [1–4]. Indeed, home NIV management has changed dramatically [5–7]. Descriptive literature based mostly on observational studies, showed that the use of long-term home NIV (LTHNIV) in RTD patients increases survival and most often avoids or postpones tracheostomy and home invasive mechanical ventilation [8]. Survey-based data are valuable resources to gain reliable data. For instance, the Eurovent survey in 2005, provided a global picture of practices regarding home mechanical ventilation (HMV) in patients with chronic respiratory failure across Europe [9]. However, since the Eurovent survey, the literature did not provide any further study on the topic. Conversely, updated information on current practice, settings, interfaces and modalities of NIV use in RTD is necessary and warranted [10-15]. Only a few studies in the literature have tried to assess settings and current NIV practices in RTD [7,16–22], the last of which is the ERS REINVENT survey [23]. It represents an international survey to collect NIV users' experience and report current clinical real-world practices for long-term home noninvasive in RTD. From these survey, we re-examined the global data obtained focusing the analysis on the Italian subsets of respondents (ITA-r), which represented the majority aiming to compare their responses to those of the non-Italian participants (non ITA-r) looking at their attitudes concerning locations and type of hospitals and units where they principally work, years of experience in NIV practice, timing and reasons for NIV initiation, modes applied and time of applications.

Materials and Methods

REINVENT was a web-based survey developed using SurveyMonkey, an online program with a cloud-based survey development application. For the purpose of the survey, the use of NIV was focused on the chronic long-term home use (LTHNIV). Patients considered were only affected by RTD, such as chest-wall deformity, neuromuscular diseases (NMD), spinal cord injury, phrenic nerve paralysis, fibrothorax post-tuberculosis, and thoracoplasty (the list was provided on the first page of the survey). Patients with restrictive parenchymal lung diseases or obesity hypoventilation syndrome were excluded since NIV has been already largely studied for both [24-26]. The aim was to deeply explore physicians' perceptions as to the use of NIV [27-29]. The European Respiratory Society (ERS) scientific committee validated the survey before its submission to ERS Assembly 2.02 (NIV dedicated group). After several revision, the ERS institutional review board approved the final version of the survey and all participants were waived to submit any consent to participate in the study. Indeed, the survey was

anonymously carried out by all participants. Only data focused on clinical experience were collected and no sensitive or other personal data. All methods were carried out in accordance with relevant guidelines and regulations.

The survey consisted of three parts. The first part included general questions about the participants' professional status and general characteristics, the type of RTD most often encountered in their hospital practice, and the personal experience with LTHNIV in RTD treatment. The second part was centered on expected clinical benefits, reasons for NIV initiation, and characteristics of ventilators used: pre-set modes, interfaces, circuits, and humidification. Ventilation modes were defined as follows: mouthpiece ventilation (MPV), spontaneous pressure support ventilation (S-PSV), spontaneous-timed PSV (ST-PSV), PSV with target volume (TV-PSV), pressure-controlled ventilation (PCV), continuous positive airway pressure (CPAP) and volume-controlled ventilation (VCV). The third and last part was referred to as "timing and type of follow-up". The full original survey is available in the supplementary material. After collecting and publishing the international data, a re-analysis of only the Italian subset of responses and a comparison to those provided during the REINVENT study was performed and described.

Statistics

Descriptive statistics analysis, including proportions, means, and standard deviation (SD) or median and interquartile range (IQR), were appropriate. A contingency table was computed, and proportions were compared using the chi-squared test. The analysis was performed with SPSS version 24. A $p<0.05$ was considered significant.

Results

Responders' characteristics. One hundred sixty-six world members of the European Respiratory Society 2.02 group focused on noninvasive ventilation responded to the international survey, whose 33 (20%) were Italian members. Italy was the most represented among the 19 Europeans and 22 non-European countries in the study. Responders were physicians (93%) and they were working mainly in community (48.5%) rather than university teaching hospital (27.3%) REINVENT international data showed community (27.3%), and university teaching hospital (64.8%; $p<0.001$) (Table 1).

Similarly to the non-ITA-r participants, in 48.3% vs 41 % of cases Italians worked mainly in a respiratory intermediate care unit (RICU). Distribution of other facilities and comparison with international data is shown in Table 2. Responders in 66.7% had similar experience in terms of years of NIV practice and use compared to non-ITA-r. LTHNIV: indications, instruments and settings. NMDs were the main RCT disease needing prescription of LTHNIV (88%) with a NMDs to chest wall disorders ratio of 9:1. Amyotrophic lateral sclerosis (ALS) was the absolute most frequent indication for LTHNIV 93% vs 78% of the international data ($p>0.5$). Figure 1 describes the ranking of reasons to start LTHNIV. Furthermore, in Figure 2, the ranking of the answers regarding the most important goals to achieve with the LTHNIV initiation is detailed. Interestingly, among all evaluations only the MIP/MEP test was considered more important for ITA-r than for international colleagues, and the difference was statistically significant ($p<0.5$) (Table 2).

The patients' preferences and feedback were similar among ITA-r and **non**–ITA-r ($p>0.5$) (Figure 3). Preferred modes of ventilations and choice of interface, circuit and type of humidification were similar among the two groups too (Figures 4 and 5) ($p>0.05$). Patients' follow-up. Patients' follow-up (Figure 6) and home care program provided did not show any significant statistical difference between NO ITA-r and ITA-r practice in terms of timing and type of offer provided ($p>0.5$).

Discussion

In this study, we re-examined the Italian subset of responses (ITA-r) extrapolated from the REINVENT study, which explored the use of LTHNIV in RTD from many countries worldwide, looking for similarities and differences with the European practice. Similarly to the international study, the vast majority of responders were respiratory physicians [94%], experts in the care of NMD patients, with more than 10 years of experience in prescribing NIV. Indeed, ITA-r were experts in the field of RCD, providing in 12% of cases with more than 50 prescriptions per year. However, while in the REINVENT study, most NO ITA-r belonged to university hospitals, the majority of ITA-r worked in community hospitals followed by university hospitals and rehabilitation hospitals. This diversification showed a significant statistical difference among the two practices ($p<0.001$). This disagreement may be explained by the location of the tertiary hub centers, with physicians engaged in the care of patients with NMD may be located more in the community/rehab hospitals than in university hospitals.

Furthermore, among the most important differences found in the REINVENT study the NO ITA-r expressed the top three perceived reasons to start LTHNIV being diurnal hypercapnia, clinical symptoms and more than 3 hospitalizations/year. Conversely, on the subset of Italian responders the three most important goals to achieve with LTHNIV in NMD patients were: first, night and day gas exchange amelioration, second, dyspnea relief and third, survival improvement. It seems like for the non-ITA-r the main focus should be the quality of life (QoL) / sleep improvement while for the ITA-r a more active targeting of the correct timing to initiate NIV to improve and counterbalance the chronic respiratory insufficiency and to prolong survival should be pursued. Given the quick and unfavorable prognosis of ALS patients and the uncertainty that LTHNIV may clearly influence it [30-32], maybe more interest should be directed towards patients' QoL and sleep amelioration than trying to precisely correct the respiratory insufficiency [33,34]. Furthermore, the MIP/MEP test was considered more important for NIV initiation for ITA-r rather than for international colleagues, and the difference was statistically significant. Reasons for this choice may be related to location of practice and availability of devices present in the hospitals considered. Among NIV modes explored, similar choices were chosen both in Europe and in Italy with the MPV the most used modality during the day only, the hybrid mode PSV-VT during the night only, and the PSV-ST one during day and night if only one mode had to be chosen for both day and night. This finding as discussed in the REINVENT study is surprising as it highlights current real practice medicine which is currently not support in the literature. Indeed, the use of hybrid modalities has not been supported in long-term home NIV and not in RCD in particular as yet [35-40]. Moreover, among ITA-r 30% of responders describes using CPAP-Auto and CPAP modes during the night These ventilation modalities do not provide effective supported ventilation and should not be used in patients with NMD, even in the presence of sleep-disordered breathing (SDB). Therefore, if they are chosen during the early stages of the NMD should be considered only temporary, and close follow-up should be warranted to quickly switch to bi-level ventilatory support. Regarding interfaces, mouthpiece and nasal pillows were the preferred ones during daytime and intuitively it may be explained for patients who need prolonged NIV and using these interfaces may still conserve the capability to eat or speak, whereas oronasal or full-face masks were the first choice overnight. This may be related to the desire to counterbalance the increased muscle weakness and augmented mouth leaks during nighttime and this was similar between the international and Italian practice [40-42]. The most frequently used circuit is the

single one associated with masks provided with exhalation holes; this is in line with recent literature evidence showing similar capability to eliminate carbon dioxide compared to double limb or single circuit with expiratory valves [43]. Heated humidification was the most frequently selected to LTHNIV, which improves the rheology of secretions that may become particularly thickened during prolonged ventilation [44]. Similar to another study focused on LTHNIV [23], Italian responders tended to adapt new patients to long-term NIV in inpatient settings, providing in most cases practical sessions with educational material for patients and caregivers, while follow-up was most often performed during outpatient visits usually every 3 months. However, the REINVENT survey was launched right before the explosion of the COVID-19 pandemic, so the global picture has already greatly changed over recent months [45], forcing clinicians to try different solutions for these frail patients. Indeed, a few randomized controlled trials have recently highlighted that adaptation to mechanical ventilation at home or in out-patient settings, rather than in hospital, is cost-effective, improves health-related quality of life and is not inferior to hospital initiation for patients with RTD [36-42,46,47]. Lastly, regarding instrumental exams scheduling and follow-up to check effectiveness of LTHNIV, similar outcomes results from the Italian experience compared to Europe, leaving the pulse oximetry the most chosen tool to sequentially monitor these patients over time. Indeed, the most important goal to achieve for Italian physicians was better night and day gas exchange amelioration, however it seems that lower than 20% of responders use PtcCO2 either every 6 months or yearly. The reason for these results may be found in the more common practice of routinely arterial blood gas analysis check, this instead of overnight ptCO2 which is not routinely performed in clinical practice due to the costs of the Co2 sensor and it's not routine availability in all ventilator machines. Moreover, there is still a lack of precise information on the presence of insurance and financial constraints on NIV prescription of different countries as recently reported; therefore, additional research is warranted [48].

The major limitations of this study are: first, the small number of responders involved; however, it was the highest percentage of responders from the international REINVENT study compared to all other countries, showing the great interest and clinical expertise on the topic among Italian physicians; second, although the survey was conducted among Italian members of the ERS assembly for NIV, specifically dedicated to noninvasive respiratory support, results may not entirely reflect the physicians' real practice and experience with long-term NIV treatment of patients; third, this study was a sub group analysis not designed a priori to collect data on

this particular topic but they were a secondary analysis. The main strengths of this study are: first, to our knowledge, only one other study focused on exploring the real clinical practice experience of the Italian respiratory physicians involved in the care of NMD patients using NIV [49]: in both studies the most common criteria to start long term NIV was daytime hypercapnia, moreover in the first study the highly specialized centers (probably the ones included in REINVENT ERS survey) used to accurately assess respiratory lung function using MIP, MEP and peak cough flow; second, although with a small number, this study delineates the practice of expert health care providers and frequent prescribers used to deal with rare ALS disease patients and NIV; third, the perception emerged from this study is that still there is still much variety of practice that needs to be explored which is very important to pave the way to further address these topics with further research.

Conclusions

In conclusion, LTHNIV has increasingly become an essential part of the NMD natural disease evolution and survival. Our data showed that based on current practice Italian respiratory specialists are frequent prescriber experts in the field of NMD patients with respiratory failure and they have similar but different approaches to the international practice. In particular, some differences emerged regarding the working location being more in community hospitals than the university hubs and about the desired targets to achieve in the NIV use in NMDs. Interestingly, the Italian focus seems more active in targeting the correct timing for NIV initiation and to correct the chronic respiratory insufficiency thus trying to prolong survival, the international vision seems more focused on the QoL experience of these patients. Further studies will be required to better detail the current Italian NIV approach to NMD patients and the post COVID19 pandemic practice changes.

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Table 1. Comparison among International participants vs Italians regarding location of work

	International	%	Italians	%	p-value
In which hospital do you work?	University	74	University	29	<0.001
	Community	17	Community	45	
	Outpatients	2	Outpatients	0	
	Private	4	Private	3	
	Rehabilitation centres	3	Rehabilitation centres	23	

Table 2. Comparison among international vs Italians regarding SNIP/MIP/MEP test for NIV initiation.

	International	%	Italians	%	p-value
MIP/MEP reduction	Extremely important	4	Extremely important	16	0.003
	Very important	19	Very important	13	
	Important	38	Important	61	
	Slightly important	29	Slightly important	7	
	Least important	10	Least important	3	

Figure 1. Reasons to start LTHNIV by Italians practitioners.

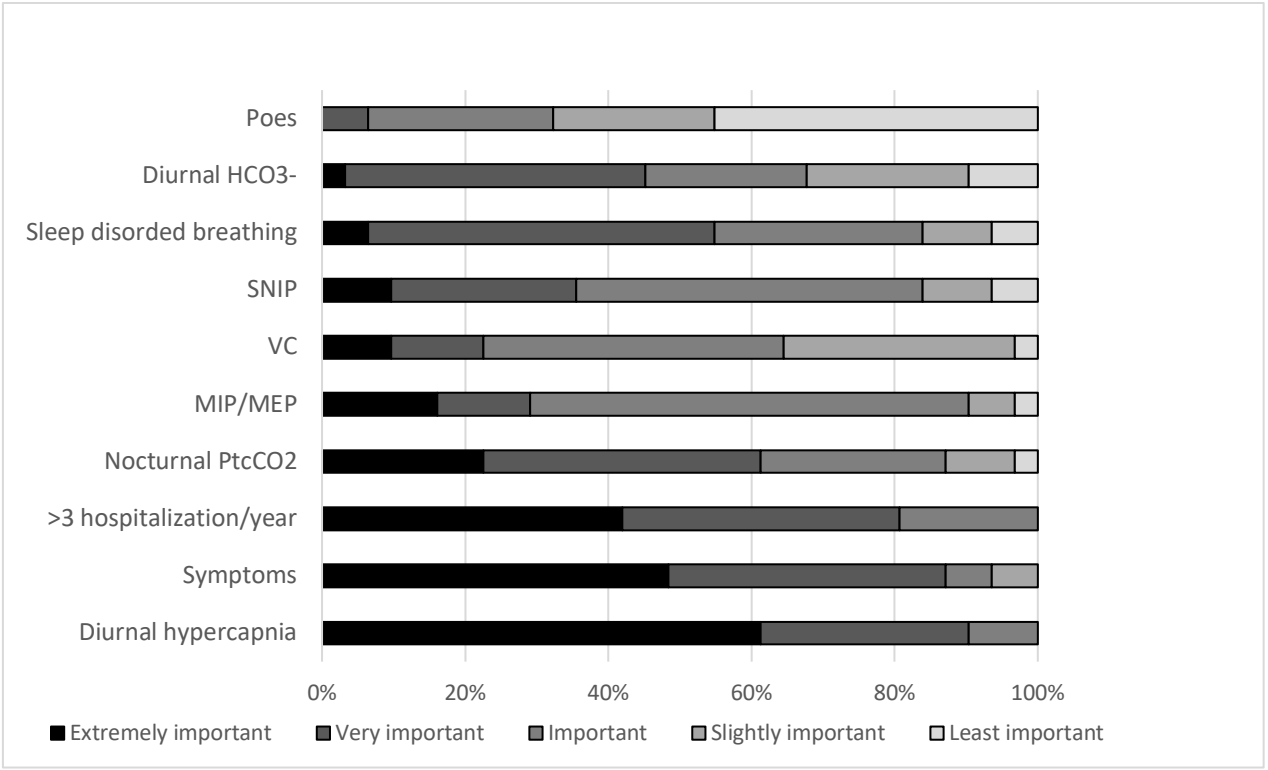

Figure 2. Ranking of the answers regarding goals to achieve with the LTHNIV by Italian practitioners.

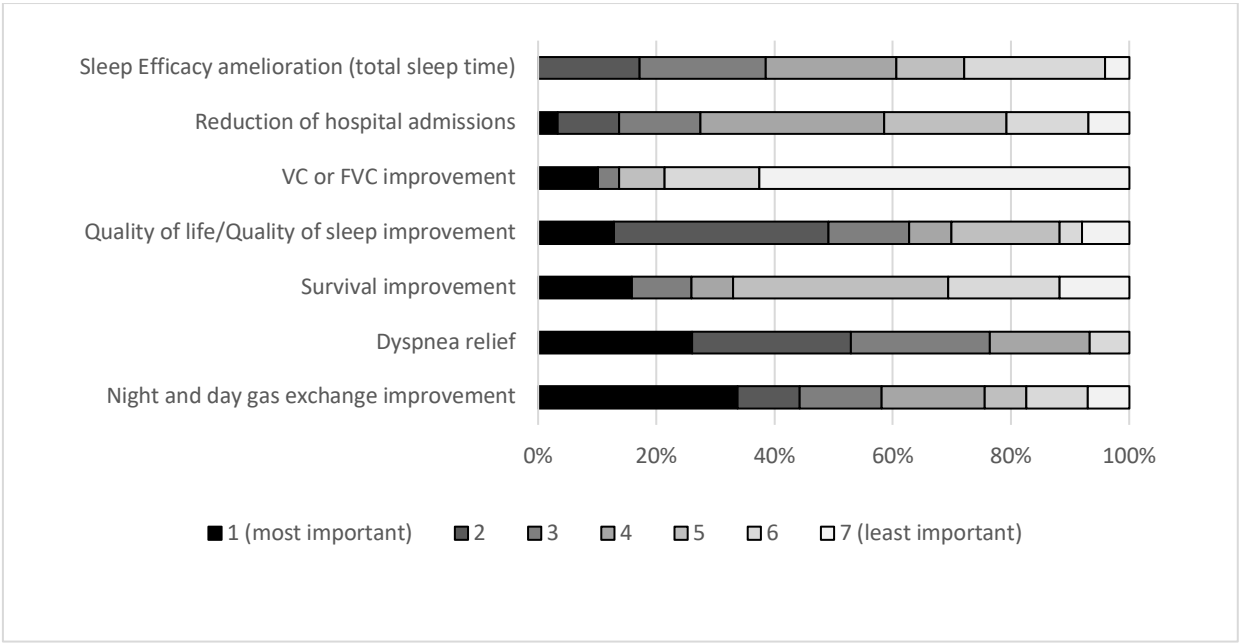

Figure 3. Criteria to choose the ventilator by Italian practitioners.

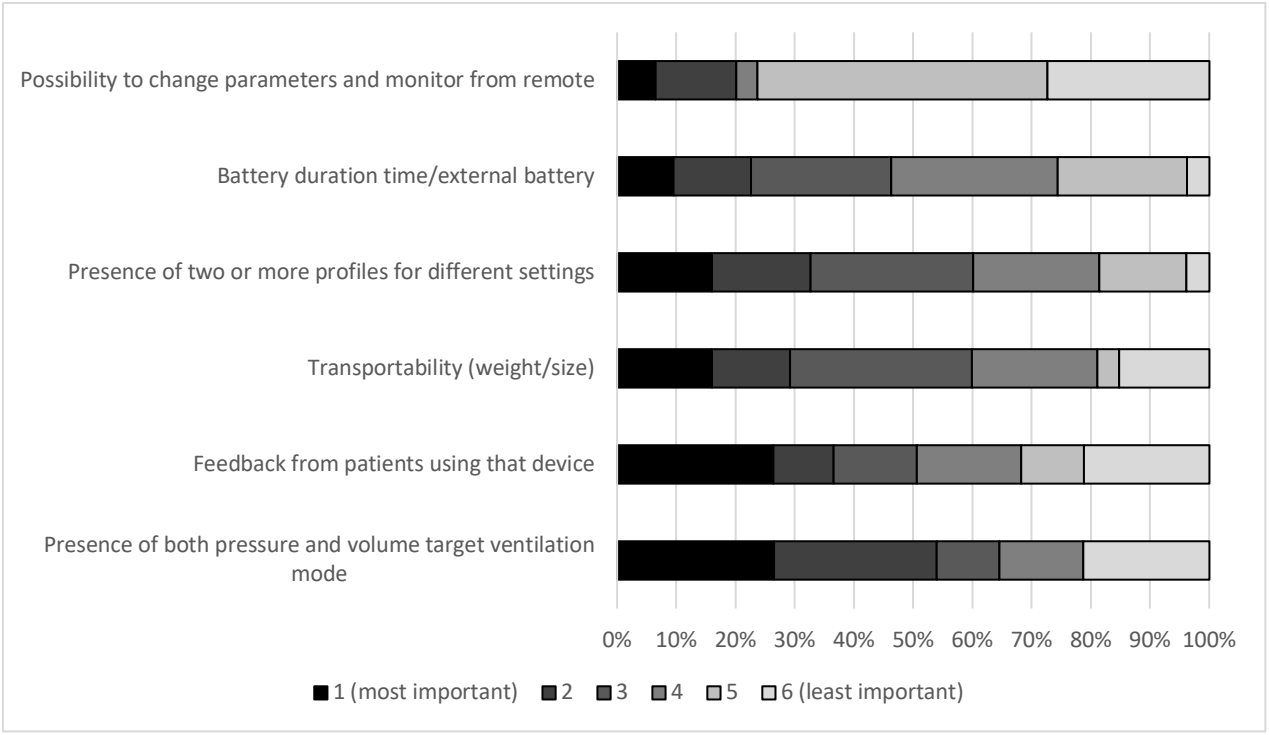

Figure 4. Ranking of ventilation modalities during day and nighttime by Italian practitioners

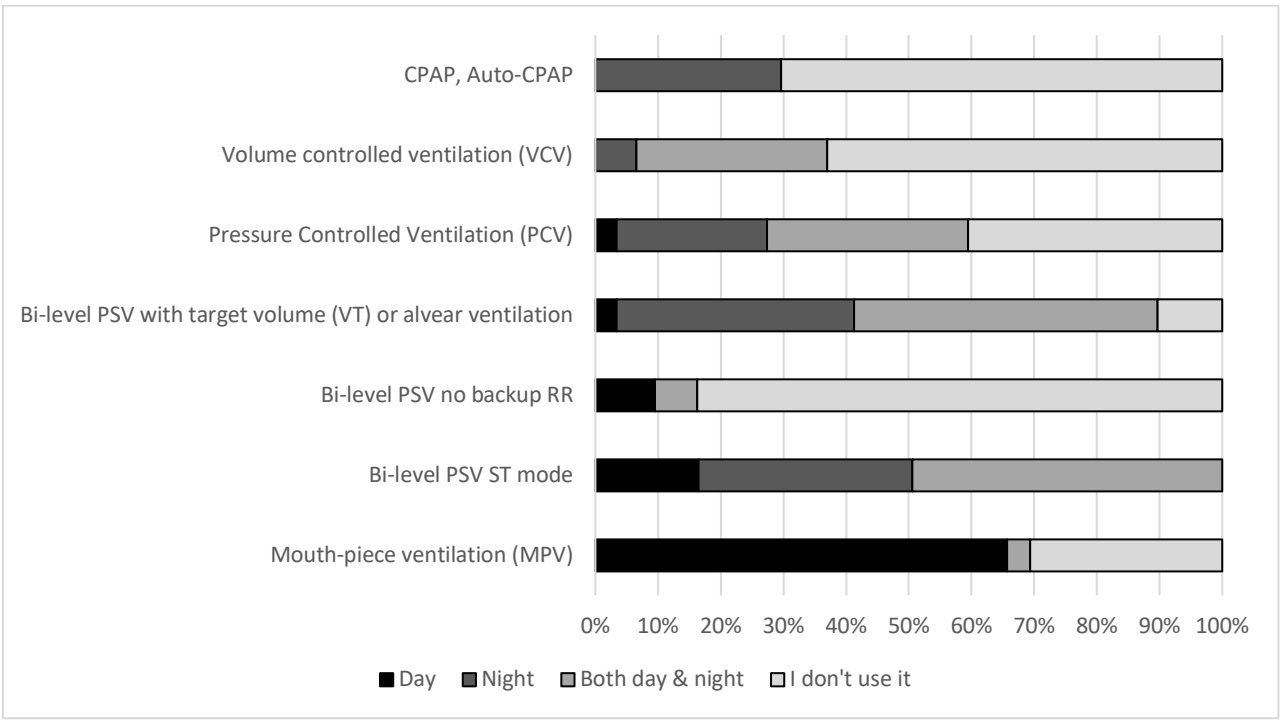

Figure 5. Interfaces selected by Italian practitioners during daytime and nighttime.

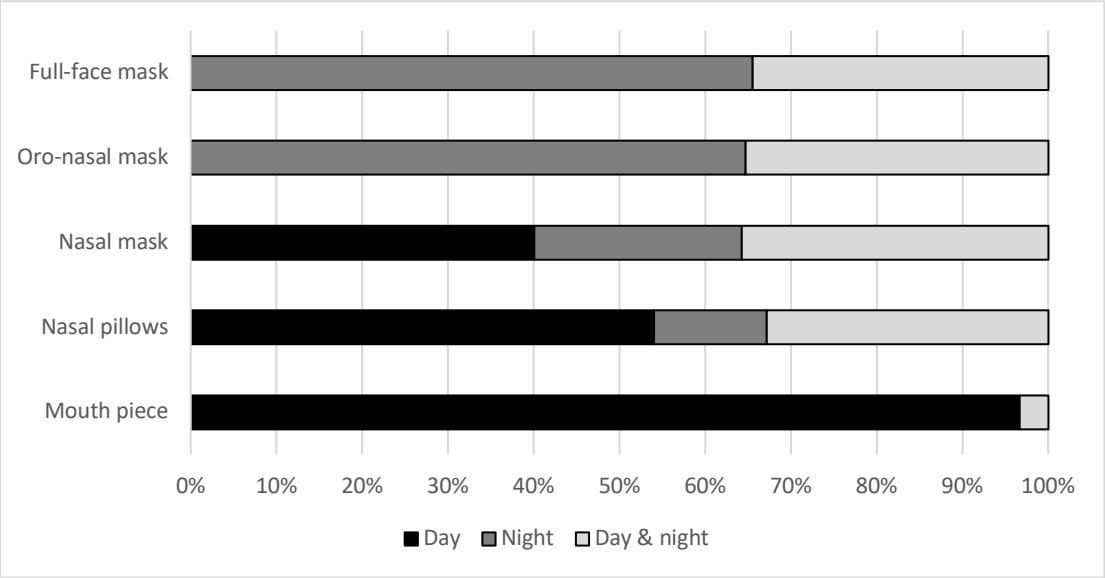

Figure 6. Type and time of follow up provided by Italian practitioners to patients in LTHNIV.