

Early View

Original article

REINVENT: International Survey on REstrictive thoracic diseases IN long term home noninvasive VENTilation

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TITLE PAGE

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REINVENT: International Survey on REstrictive thoracic diseases IN long term home noninvasive VENTilation

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Key words: Noninvasive ventilation, chronic respiratory failure, restrictive thoracic disease, neuromuscular disease, chest wall disorders, international survey.

Abstract:

Background and aim: Little is known about the current use of long-term home non-invasive ventilation (LTHNIV) in restrictive thoracic diseases (RTD), including chest wall and neuromuscular disorders (CWD, NMD). This study aimed to capture the pattern of LTHNIV in RTD patients via a web-based international survey.

Methods: The survey involved ERS Assembly 2.02 (NIV dedicated group) October-December 2019.

Results: 166/748 (22.2%) members from 41 countries responded; 80% were physicians, of whom 43% worked in a respiratory intermediate intensive care unit (RIICU). The NMD:CWD ratio was 5:1, Amyotrophic lateral sclerosis (ALS) being the most frequent indication within NMD (78%). The main reason to initiate LTHNIV was diurnal hypercapnia (71%). Quality of life/sleep was the most important goal to achieve. In 25% of cases, clinicians based their choice of the ventilator on patients' feedback. Among NIV-modes, pressure support ventilation spontaneous-timed (PSV-ST) was the most frequently prescribed for day and night-time. Mouthpieces were the preferred daytime NIV interface, whereas oro-nasal masks the first choice overnight. Heated humidification was frequently added to LTHNIV (72%). Single-limb circuits with intentional leaks (79%) were the most frequently prescribed. Follow-up was most often provided in an outpatient setting.

Conclusions: This ERS survey illustrates physicians' practices of LTHNIV in RTD patients. NMD and, specifically, ALS were the main indications for LTHNIV. NIV was started mostly because of diurnal hypoventilation with a primary goal of patient-centred benefits. Bi-level PSV-ST and oro-nasal masks were more likely to be chosen for providing NIV. LTHNIV efficacy was assessed mainly in an outpatient setting.

Short title: REINVENT international survey of NIV in RTD

Introduction

In the mid 1980's, non-invasive ventilation (NIV) became the reference treatment for chronic respiratory failure (CRF). This allowed a drastic decrease in invasive ventilation (tracheostomy), while improving survival, quality of sleep and quality of life (QoL) in patients with both obstructive and restrictive thoracic disorders (RTD)[1-4]. Over the last decades, the management of home NIV has changed dramatically. [5-7]

Published data support the use of NIV in RTD patients and have shown its clinical and physiological benefits. Long term NIV increases the likelihood of prolonged survival and thus most often allows avoiding tracheostomy and home invasive mechanical ventilation. Except for a few RCT in Amyotrophic lateral sclerosis (ALS)[8], the positive impact of NIV in RTD is based mainly on observational uncontrolled studies. Survey-based findings represent a valuable alternative source of data. They can reliably describe the practices of clinicians in different domains of medicine, such as home non-invasive ventilation.

In 2005, the Eurovent survey provided a comprehensive picture of practices regarding home mechanical ventilation (HMV) in patients with CRF across 16 European countries. [9] For the first time, it identified patterns of use and settings of HMV across Europe. This study helped many physicians to confront their clinical practice to the information gathered.

Since the Eurovent survey, there has been no follow-up study of the use of NIV in patients affected by RTD across Europe. Indeed, there is an evident gap in the literature on this topic. Updating information on settings, interfaces and modalities of NIV use in Europe (i.e. mouthpiece ventilation) is necessary and warranted.[10-14] To the best of our knowledge, only a few studies in the literature tried to assess settings and current NIV practices in RTD [7, 15-19].

We therefore performed an international survey to collect NIV users' experience and report the current clinical real-world practices for REstrictive disorders IN long term home noninvasive VENTilation: REINVENT.

Material and methods

Survey development

This web-based survey was developed using Survey-Monkey, an online platform with a cloud-based survey development application.

The ideated survey was then conducted to explore physicians' clinical management of long-term NIV in the treatment of CRF due to RTD. For the purpose of the survey, "long-term home, non-invasive ventilation" (LTHNIV) included only patients with RTD as defined hereafter. Respiratory diseases included were: chest-wall deformity (CWD), neuromuscular diseases (NMD), spinal cord injury, phrenic nerve paralysis, fibrothorax-post TB, and thoracoplasty. We excluded patients with obesity hypoventilation syndrome (OHS) or parenchymal restrictive lung diseases (RLD). A list of illnesses included was provided on the first page of the survey with a brief explanation of the aim of our research.

The survey was developed based on previous work, exploring physicians' perceptions as to use of NIV [20-22]. The survey instrument was designed after a thorough literature review to generate relevant survey items. A panel of ERS experts on NIV, part of the Steering Committee of the project, reviewed the survey items for content validity, relevance, and ability to discriminate among respondents. The ERS management group then revised the survey by adding further details based on previous proposed survey studies. The final survey questionnaire included various formats such as Likert scales, ranking, and yes/no, but did not allow for open-ended questions.

The survey consisted of three parts. The first part included general questions about the participants' professional status, general characteristics, experience with LTHNIV in the treatment of RTD and the type of RTD most often encountered in their hospital practice. The second part was mainly centred on reasons for NIV initiation, clinical benefits expected, and characteristics of ventilators used: pre-set modes, circuits, interfaces, and humidification. Ventilation pre-set 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), volume controlled ventilation (VCV). The third and last part was referred to as "timing and type of follow-up". The full survey is in the supplement material.

Survey testing

We administered the survey to 10 respondents, including pulmonologists and critical care physicians, to test the comprehensiveness, clarity, and validity. We estimated interrater reliability using Cohen's kappa test with a threshold value above 0.4 (i.e. moderate agreement).

Survey administration

To identify clinicians interested and involved in NIV practices, we contacted the members of ERS assembly 2, Group 2.02. This group is a heterogeneous, multidisciplinary and multi-professional group, incorporating physicians with different educational backgrounds, such as pulmonology, anaesthesiology, internal medicine, intensive care medicine and emergency medicine, as well as allied healthcare professions (nurses and physiotherapists). These professional figures participate together in this group based on their common interests and expertise in NIV practice.

Email notifications with a link to a web-based questionnaire were sent in September 2019 to all the 748 members of ERS Assembly 2, Group 2.02. Reminders were sent every four weeks. The survey was closed in December 2019.

Data entry and analysis

We reported descriptive statistics, including proportions, means, and standard deviation (SD) or median and interquartile range (IQR), when appropriate.

The respondents were grouped based on the type of ward in which they principally worked: 1) critical care (emergency department, ICU, pulmonary ward + high dependency unit), and 2) general (pulmonary ward & general ward vs. rehab, private practice, outpatient clinic) to allow comparisons. Contingency tables were computed, and proportions were compared using the chi-squared test. Analyses were performed with SPSS version 24. A p value <0.05 was considered significant.

Results

Survey response rate

Out of the 748 ERS Assembly 2.02 members, 166 health care professionals responded to the survey, i.e. a response rate of 22.1%. Respondents belonged to 41 different countries: 19 European and 22 non-European countries. The full list is included in an e-table (on-line supplement).

Respondents' characteristics

The majority of responders worked in university teaching hospitals (64.8%), followed by community hospitals (22.4%), rehabilitation centers (6.7%), private hospitals/clinics (4.2%), and outpatient clinics (1.8%). They worked mainly in respiratory wards with a dedicated respiratory intermediate intensive care unit (RIICU) (43%). Distribution of other facilities is shown in Table 1. Responders belonged to different health care professions: physicians, physiotherapists, nurses, and others. The most represented were physicians (80%). They were generally very experienced in NIV, most of them having more than 10 years of experience (61%). Fifteen percent of respondents reported more than 50 ventilator prescriptions per year.

Indication for LTHNIV

The NMD was the most important indication for 80.9% of the respondents, with ALS representing 78% of these cases, followed by Duchenne Muscular Dystrophy (DMD, 11%), CWD 14.5% and others (4.6%).

The most important reasons for initiating LTHNIV reported by respondents are shown in figure 1. Respondents ranked the most significant treatment targets to achieve using LTHNIV, as shown in figure 2.

Instruments and Settings

Ventilators and settings

Participants were more likely to choose a specific ventilator based on the following characteristics of devices: good feedback from patients (25%), presence of both pressure and volume ventilation options, and transportability (20%). (Figure 3).

The majority of interviewees used MPV during daytime only (65%), followed by S-PSV (17%), and ST-PSV (9%). MPV was more frequently used in general respiratory wards (rehabilitation and acute wards) than in critical care wards (ICU, RICU, ED) ($p=0.015$). Respondents reported that TV-PSV (29%) was the preferred mode of ventilation during night-time, followed by PCV (20%), ST-PSV (19%), CPAP (18%), and VCV (10%). However, if one mode was chosen for both day and night-time ventilation, then ST-PSV was the most frequently used (36%), followed by TV-PSV (25%), PCV (16%), and VCV (15%). All modalities used between day and night time are detailed in figure 4.

Clinicians were more likely to use a single-limb circuits with intentional leaks or exhalation ports (79%), followed by expiration valves (13%) and double-limb circuits (8%).

Interfaces preferred by all responders during the day and night-time ventilation are shown in figure 5.

No further significant results were found from other comparisons among groups considered. Clinicians working both in general respiratory wards and critical care wards reported similar preferences in terms of modes of ventilation, interfaces and circuits' configuration.

Most prescribers added humidification to NIV. A heated humidifier was the first choice (72%) followed by heat and moisture exchangers (HME) (20%). No humidification was prescribed in only 8% of cases.

Patients' follow-up

The vast majority of responders initiated patients to NIV as inpatients (67%). During the hospital stay, an educational program was provided for new patients via either educational material combined with practical sessions for patient and caregiver (40%) or only practical sessions for patient and caregiver (33%). Lack of educational programs was reported by 27% of respondents. The prescriber was usually also involved in long term patient follow-up (95%). Follow-up was performed during outpatient visits in 65% of cases, as inpatients in 18% and at home in 12%. The remaining 5% of responders described a combined schedule of outpatient visit and telemedicine or ventilator tele-monitoring. The different types of follow-up provided are reported in figure 6. Finally, on top of follow up visits described, in 65% of cases, a home care program with control visits was provided through either a physician (11%), a nurse (14%), a physiotherapist (11%), or a home care provider (19%). The remaining responders (10%) provided a home care program combining at home follow-up visit by health care practitioners (i.e. physician, nurse, physiotherapist) and telemedicine or tele-monitoring. Conversely, in 35% of cases, the option of a home care program was not available for RTD patients on LTHNIV.

Discussion

The REINVENT survey study explored the clinicians' perspectives as to use of LTHNIV for RTD among professionals of different countries and members of the ERS assembly group on non-invasive respiratory support. Fifteen years after EUROVENT, this study describes the type of RTD patients requiring NIV, settings, modes, and interfaces used. Survey response rate was 22.2%, which is in line with reported response rates of internet-based surveys [23]. Responders were mainly physicians (80%) involved in the care of NMD patients, primarily in teaching hospitals (64.8%) and community hospitals (22,4%). This is in line with what was previously reported in EUROVENT [9]. The vast majority of responders (61%) had extensive experience in LTHNIV (>10 years). Therefore this study provides an indicative picture of experienced physicians working in the field of RTD.

The most important reasons for LTHNIV initiation were diurnal hypercapnia, hospitalizations for respiratory failure, muscle weakness symptoms, and nocturnal hypercapnia. Interestingly, having an FVC lower than 80% of predicted, which is recommended for NIV initiation in current ALS treatment guidelines, was not in the top three answers [24]. Targets of LTHNIV treatment were more often related to QoL and quality of sleep than to increasing survival. Most probably, a lesser importance is given to survival in RTD patients and, in particular, in ALS patients, given their prognosis, which seems to be only partially influenced by LTHNIV [25-27]. More importance is given to the quality of patient's experience during LTHNIV used during day and night-time [28]. Indeed, less evidence is present in the literature for LTHNIV in RTD use compared to other respiratory diseases such as COPD, where LTHNIV management has only recently been defined [29].

Interestingly, prescribers took into high consideration feedbacks from other patients already using the same machines. Indeed, these feedbacks were used in 25% of cases to drive the choice of the ventilator. Secondly, the presence of both pressure and volume modes was important: many prescribers chose hybrid modes with pressure and volume settings in RTD patients. Thirdly, transportability and battery autonomy or presence of an external battery were on the top of the list of aspects to consider when choosing a ventilator. Given the weakness and total dependence of NMD patients, these items play a vital role in patient autonomy. Patients may feel safe despite depending on the ventilator, and they may continue to enjoy a good quality of life moving around with their caregivers and outside their homes.

Ventilation modes with combined pressure and volume settings (hybrid modes: TV- PSV), are often chosen by the prescribers to be preferably used overnight. These results are interesting and deserve to be reviewed in depth. Indeed, hybrid modes are relatively new setting modalities in the LTHNIV scenario, and therefore their potential is yet to be explored [30]. These modes combine pressure and volume modes by delivering a targeted volume via a predefined pressure range (minimal and maximal IPAP) set on the ventilator[31]. There is no support in the literature for the use of these modes in long term NIV. Therefore the question is why opt for them when simple bi-level PSV/ST modality is widely used by prescribers and accepted by the patients. To date, only a few studies have explored the use of these hybrid modes in patients with OHS and chronic respiratory failure [32-34]. Although, intuitively in patients with RTD, hybrid modes could present several advantages, there is no evidence as

yet in the literature as to their real effectiveness. For instance, their response in presence of leaks may be unpredictable in NMD patients. Since many ventilators already provide these modes, and prescribers use them for the treatment of RTD patients as confirmed in this survey, studies are warranted to explore their benefit (or absence of) in more detail in this population. However, it is important to highlight that despite the larger use of hybrid modes, PSV-ST was the preferred mode for both day and night-time ventilation. This confirms the large knowledge and practicability of this mode among prescribers surveyed.

The extensive use of CPAP/auto-CPAP at night in RTD patients highlighted by this survey deserves a comment. These modalities are not a ventilation mode, and they are not indicated in RTD albeit in the presence of sleep-related disordered breathing (SDB). SDB may be one of the presenting symptoms in NMD patients. When using CPAP/auto-CPAP in NMD, a close follow up is warranted to switch to bi-level support when required, and provide adequate respiratory support.

The most frequently used circuit is a single-limb tubing with an intentional leak port. This is in line with recent evidence describing this option as easier to use, and allowing a sufficient CO₂ exhalation compared to single or double limb circuits with expiratory valves [35]

In this survey, in NMD (mostly ALS), mouthpiece ventilation is the most frequently used mode of daytime ventilation. Although nasal masks and nasal pillows are used less than mouthpieces during daytime ventilation only, when patients require prolonged ventilation during the day and night time, they become the first choice. Oro-nasal interfaces are used preferably for nocturnal ventilation only, probably to avoid excessive mouth leaks [18].

Humidification seems to be very important for prescribers who prefer active humidification (72% of cases) to HME (20% of cases). Indeed, bronchial secretions may become very thick during prolonged ventilation; therefore, heated humidification plays a role in improving the rheology of secretions and overall quality of ventilation [36].

Follow-up was mainly provided by physicians via outpatient visits or via a home care provider. At the time of the survey a very large number of prescribers did not provide any home care program. The picture has changed over the last months[37]. Indeed, a few RCTs have recently highlighted that initiation of mechanical ventilation at home is cost effective, improves HRQL and is not inferior to hospital initiation also for patients with RTD [38-42]. This survey was launched and concluded right before the COVID-19 pandemic, which had a profound impact on our way of managing LTHNIV patients. Social distancing has promoted the use of tele-monitoring, telecommunication and tele-visiting within many specialties and many patients. The scenario ahead of us will probably be very different: it may be preferable for these vulnerable patients to be managed by remote monitoring. A further upgrade to improve control and to modify ventilator settings remotely will probably be provided.

This study has a few limitations. First, although the survey was conducted among members of the ERS assembly for NIV, specifically dedicated to non-invasive respiratory support, results may not be representative of physicians' clinical experience and perceptions with long term

NIV treatment of patients with RTD across Europe. Secondly, the 22% response rate could be considered as relatively low. It is however in line with reported response rates for electronic surveys. Also, some RTDs are rare diseases sometimes managed only in few dedicated and specialized centres: this could have further affected the overall response rate. Thirdly, there may be a selection bias in this survey, with almost 2/3 of responders working in university teaching hospitals: this may impact on severity of cases, and choice of devices and settings.

The major strengths of our study are that it was the first study since Eurovent that focused on LTHNIV in RTD. Secondly, respondents could not skip sessions or answers: therefore full data collection of respondents' opinions was guaranteed. Lastly, we gathered quality responses from health-care professionals who are experts in this field.

In conclusion, the present REINVENT survey has provided a global picture of LTHNIV in patients with RTD and, in particular, with rare NMD who require long term ventilation. Compared to the previous EUROVENT survey, it showed different reasons to initiate LTHNIV and goals to be achieved. Patterns of ventilation and modes used have considerably changed, highlighting the increase in use of combined modes in LTHNIV. Follow-up of these patients before the COVID-19 pandemic was mainly via out-patient visits or home care programs with an in-person appointment. We expect that after the COVID 19 global pandemic, common practices will change significantly via the use of tele-monitoring and telehealth techniques. Further studies are needed to evaluate what has changed among the clinical practice of LTHNIV in these patients affected by rare NMD.

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Fig 1 Reasons to start LTHNIV in RCD

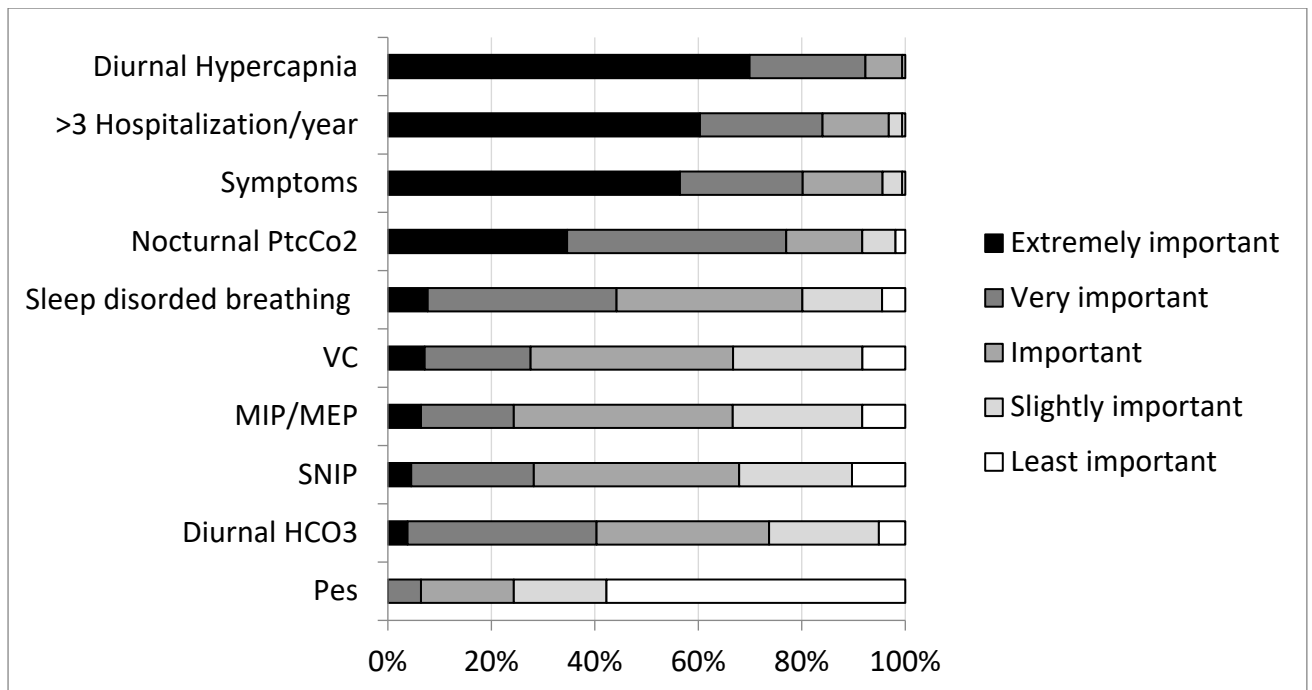


Fig2 Treatment's targets of LTHNIV for RCD

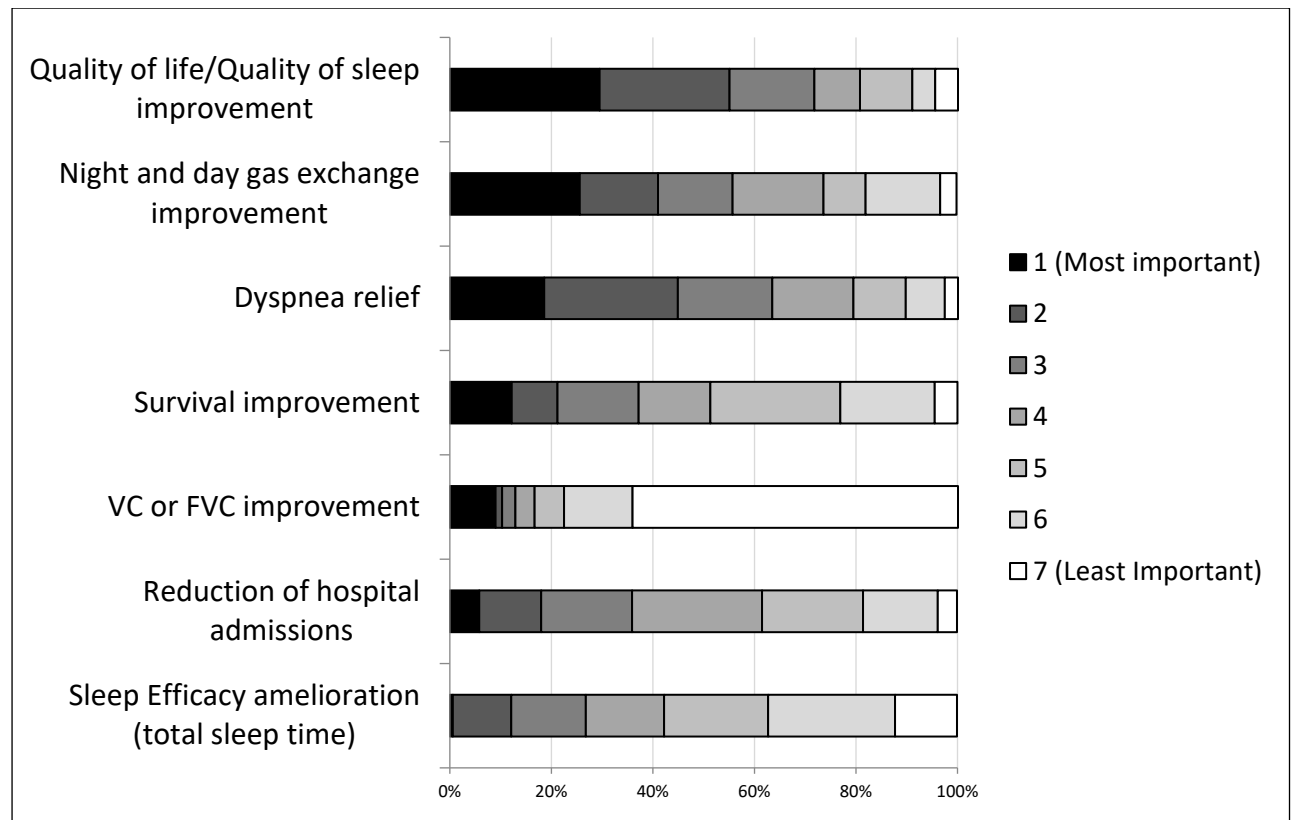


Fig 3 Choice of ventilator

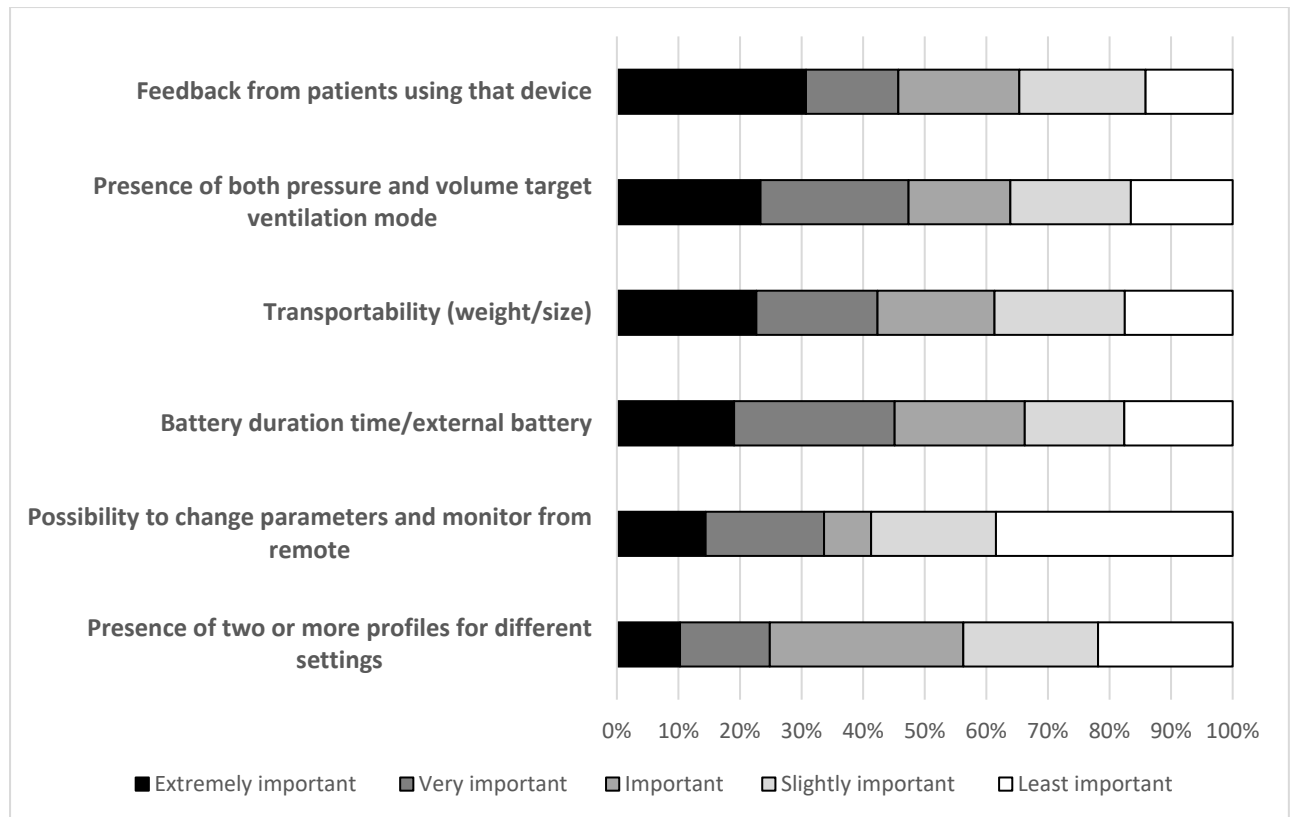


Fig 4 Modes of Ventilation

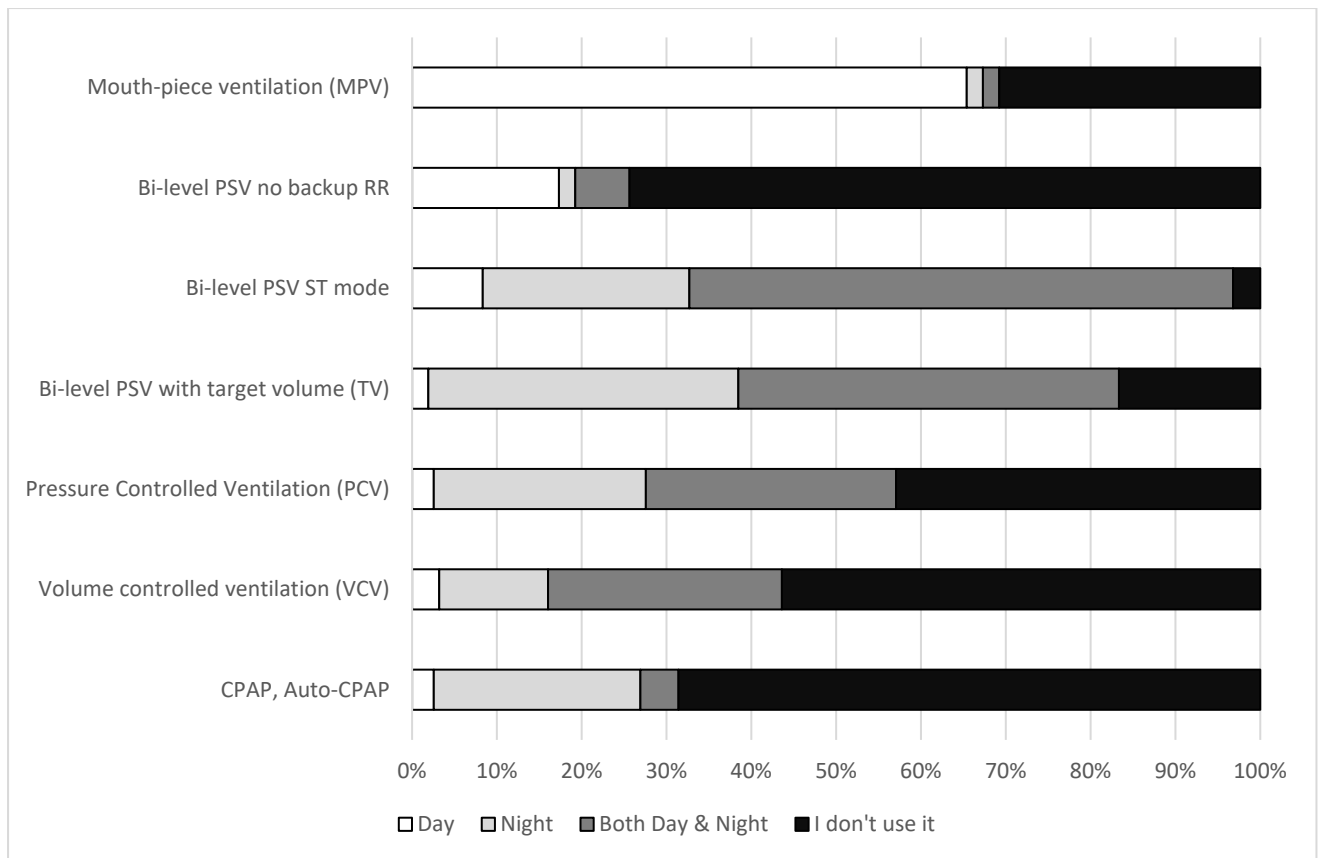


Fig 5 Interfaces used during LTHNIV

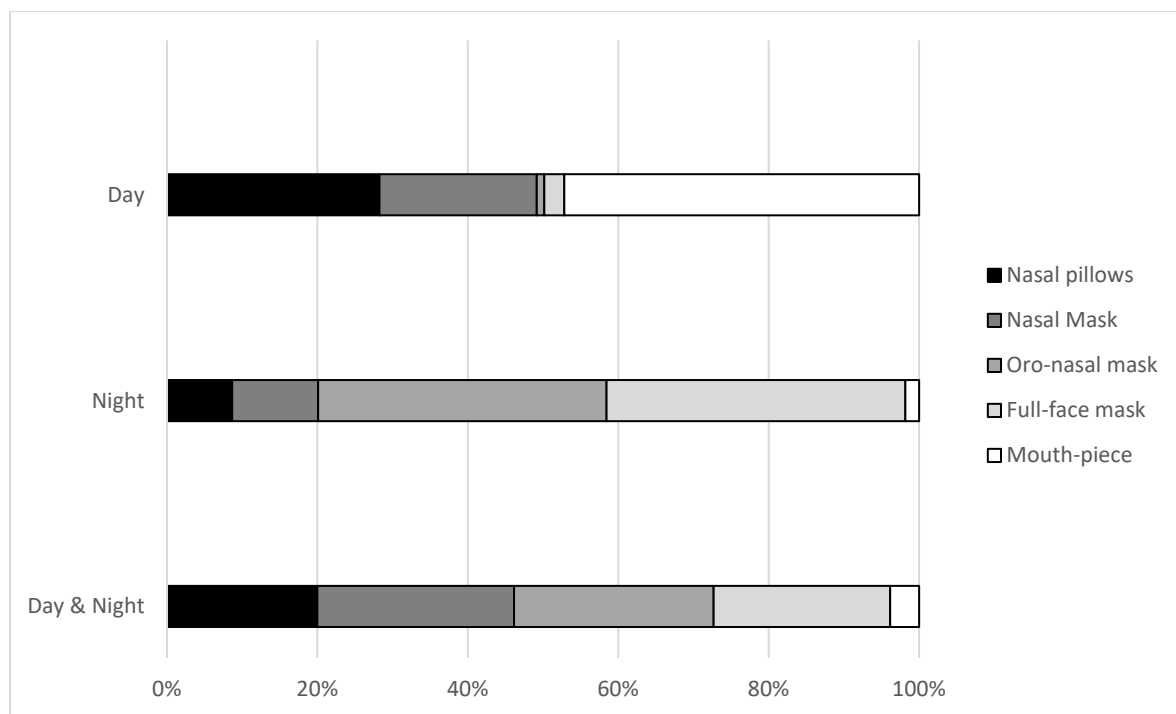
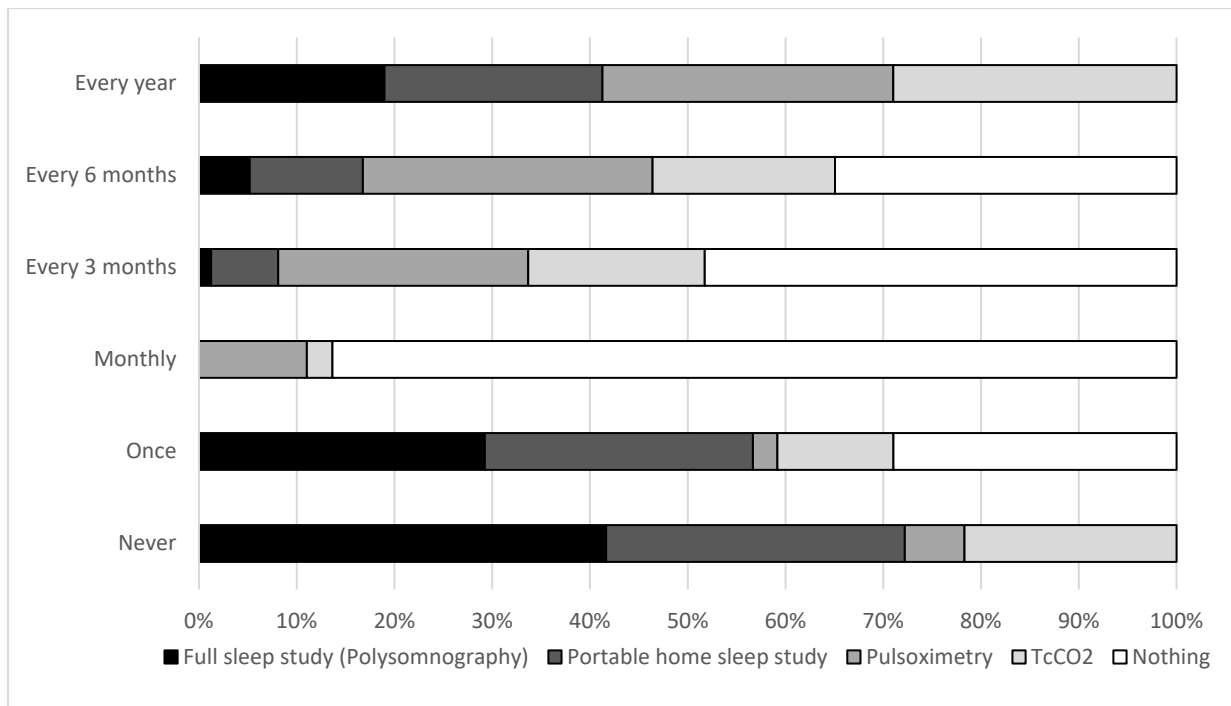


Fig 6 Follow up provided for LTHNIV



Supplement E- Table. Survey Response rate

Countries	N	%
Italy	33	19.88%
United Kingdom	19	11.45%
Spain	16	9.64%
Germany	11	6.63%
Portugal	10	6.02%
Netherlands	8	4.82%
France	7	4.22%
Poland	6	3.61%
Switzerland	5	3.01%
Australia	4	2.41%
Canada	3	1.81%
Greece	3	1.81%
Malaysia	3	1.81%
Turkey	3	1.81%
Austria	2	1.20%
Chile	2	1.20%
Croatia	2	1.20%
Estonia	2	1.20%
Ireland	2	1.20%
Pakistan	2	1.20%
Romania	2	1.20%
Russia	2	1.20%
Albania	1	0.60%
Argentina	1	0.60%
Czech Republic	1	0.60%
Finland	1	0.60%
Hungary	1	0.60%
Kazakhstan	1	0.60%
Kuwait	1	0.60%
Lithuania	1	0.60%
Mexico	1	0.60%
Norway	1	0.60%
People 's Republic of China	1	0.60%
Peru	1	0.60%
Serbia	1	0.60%
Singapore	1	0.60%
Slovenia	1	0.60%
Sri Lanka	1	0.60%
Sweden	1	0.60%
Tunisia	1	0.60%
Venezuela	1	0.60%
Total	166	100.00%

Table Legend: The highest response rates/country (\geq than 10 responses per country) came from Italy, UK, Spain, Germany, and Portugal. Intermediate response rates/country (between 5 and 10

answers) were provided by France, The Netherlands, Poland, and Switzerland. The remaining countries involved provide low response rates/country (<5 responses).

1. Survey of long-term home NIV (LTHNIV) in patients with restrictive chest disorders (RCD)

Thanks for taking part in this survey.

Before you start:

This survey aims to have an overview on the use of long-term home non-invasive ventilation (LTHNIV) in patients with restrictive chest disorders (RCD).

The results will be used to know the current clinical practice of long-term NIV use in this specific field.

Analysis of the gathered data will be performed and research results published.

Please feel free to leave your email at the end of the survey to be informed on the results.

The following list includes the restrictive chest disorders (RCD) considered in this survey:

- Chest-wall deformity
- Neuromuscular disease
- Spinal cord injury
- Phrenic nerve paralysis
- Fibrothorax-post TB thoracoplasty

Please note that Obesity Hypoventilation Syndrome (OHS) and Restrictive Lung Diseases (RLD) WILL NOT BE included in the present survey.

Thanks for your cooperation!

Time from start to finish: 10 minutes.

* 1. In which country do you work?

* 2. In which type of hospital do you work?

- ☐ University Teaching Hospital
- ☐ Community Hospital
- ☐ Private hospital/Clinic
- ☐ Outpatient Clinic
- ☐ Rehabilitation centre

* 3. In which type of unit do you work?

* 4. How many years of experience do you have with NIV, approximately?

- ☐ <1
- ☐ 1-5
- ☐ 6-10
- ☐ >10

* 5. Which is your professional status?

- ☐ Physician
- ☐ Nurse
- ☐ Respiratory therapist
- ☐ Physioterapist
- ☐ Others

* 6. On average, how many new prescriptions of long-term NIV did you perform for patients with Restrictive Chest Disorders during the last 12 months?

- ☐ 0
- ☐ 1-10
- ☐ 11-25
- ☐ 26-50
- ☐ >51

2.

* 7. Over the last 12 months, which is the Restrictive Chest Disorder you most prescribed long-term NIV for?

- ☐ Chest-wall deformity
- ☐ Neuromuscular disease, Phrenic nerve paralysis, Spinal cord injury
- ☐ Fibrothorax-post TB thoracoplasty
- ☐ Other (please specify)

3.

* 8. Which of the following neuromuscular disease you see the most? Please select one.

- | | |
|---|---|
| <input type="radio"/> Muscular dystrophy | <input type="radio"/> Myopathy |
| <input type="radio"/> Myotonic dystrophy | <input type="radio"/> Spinal muscular atrophy |
| <input type="radio"/> Amyotrophic lateral sclerosis (ALS) | <input type="radio"/> Spinal cord injury |
| <input type="radio"/> Multiple sclerosis | <input type="radio"/> Phrenic nerve paralysis |
| <input type="radio"/> Myasthenia gravis | |

4.

Please remember to answer all the questions thinking of the restrictive chest disease (RCD) you see the most in Clinic.

* 9. Which of the following factors do you consider more important to initiate long-term NIV in these patients? Please rate from extremely important to least important.

	Extremely important	Very important	Important	Slightly important	Least important
Symptoms of muscles weakness (orthopnoea, dyspnea, use of accessory muscles)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
VC or FVC < 80% pred	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
SNIP < 40 cmH2O or 3.9kPa	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
MIP/MEP reduction	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Esophageal pressure (Pes) or transdiaphragmatic pressure (Pdi) measurements	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Sleep disordered breathing (AHI >5)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Diurnal HCO ₃ ⁻ >26 mmol/L with normal PaCO ₂	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Increase nocturnal PaCO ₂ /PtCO ₂	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Diurnal hypercapnia PaCO ₂ >45mmhg	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
>3 hospitalization/year for respiratory failure	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

* 10. Which of the following clinical benefits do you expect to achieve prescribing long-term NIV in these patients?

[1= The most important; 7= The least important]

Note: you can rank your answer inserting the number or sliding the bar.

N/A= not applicable.

<input type="text"/>	VC or FVC improvement	<input type="checkbox"/> N/A
<input type="text"/>	Night and day gas exchange improvement	<input type="checkbox"/> N/A
<input type="text"/>	Dyspnea relief	<input type="checkbox"/> N/A
<input type="text"/>	Quality of life/Quality of sleep improvement	<input type="checkbox"/> N/A
<input type="text"/>	Reduction of hospital admissions	<input type="checkbox"/> N/A
<input type="text"/>	Survival improvement	<input type="checkbox"/> N/A
<input type="text"/>	Sleep Efficacy amelioration (total sleep time)	<input type="checkbox"/> N/A

* 11. How do you rank the importance of the following NIV device characteristics when choosing a machine for long-term NIV in these patients?

[1= The most important; 6= The least Important]

Note: you can rank your answer inserting the number or sliding the bar.

N/A= not applicable.

<input type="text"/>	Transportability (weight/size)	<input type="checkbox"/> N/A
<input type="text"/>	Battery duration time/external battery	<input type="checkbox"/> N/A
<input type="text"/>	Presence of both pressure and volume target ventilation mode	<input type="checkbox"/> N/A
<input type="text"/>	Presence of two or more profiles for different settings	<input type="checkbox"/> N/A
<input type="text"/>	Possibility to change parameters and monitor from remote	<input type="checkbox"/> N/A
<input type="text"/>	Feedback from patients using that device	<input type="checkbox"/> N/A

* 12. Which ventilation mode do you prefer to use during day and night for long-term NIV in these patients?

	Day	Night	Both Day & Night	I don't use it
Bi-level PSV no backup RR	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Bi-level PSV ST mode	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Bi-level PSV with target volume (VT) or alveolar ventilation	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Volume controlled ventilation (VCV)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Pressure Controlled Ventilation (PCV)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
CPAP, Auto-CPAP	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Mouth-piece ventilation (MPV)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

* 13. Which interface do you usually prefer for long-term NIV in these patients?

	Day	Night	Day & Night
Nasal Mask	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Nasal pillows	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Oro-nasal mask	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Full-face mask	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Mouth-piece	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

6.

* 14. What type of **CIRCUIT** do you usually prescribe for long-term NIV in these patients?

Please rate the answer (1= most used, 4= least used).

Note: you can rank your answer inserting the number or sliding the bar.

N/A= not applicable.

<input type="text"/>	Double circuit	<input type="checkbox"/> N/A
<input type="text"/>	Single circuit including a true expiratory valve	<input type="checkbox"/> N/A
<input type="text"/>	Single circuit including intentional leaks (Whisper swivel, or plateau valve)	<input type="checkbox"/> N/A
<input type="text"/>	Single circuit including intentional leaks (Exhalation holes built-in the mask)	<input type="checkbox"/> N/A

* 15. Which type of **HUMIDIFIER** do you use while prescribing long-term NIV for Restrictive Chest Disorders?

- ☐ None
- ☐ Heated Humidifier
- ☐ Heat and Moisture Exchanger (HME)

* 16. In your practice, how do you most commonly adapt a new patient with Restrictive Chest Disorders to long-term NIV?

- ☐ Out patient
- ☐ In patient

7.

* 17. Do you have any educational program after NIV adaptation for Restrictive Chest Disorders patients in long-term NIV?

- ☐ Yes, educational material and practical sessions for patient and caregiver
- ☐ Yes, practical sessions for patient and caregiver
- ☐ No, we do not have any educational program

8.

18. Are you involved in the follow-up of long-term ventilated patients with Restrictive Chest Disorders?

- ☐ Yes
- ☐ No

9.

* 19. Where do you usually follow-up patients with Restrictive Chest Disorders on long-term NIV adaptation?

- ☐ Home follow-up
- ☐ Out-patient follow-up
- ☐ In-patient follow-up
- ☐ Other (please specify)

* 20. On average, how often do you schedule the **follow-up visits** for patients with Restrictive Chest Disorders on long-term NIV?

- ☐ Every month
 ☐ Every year
☐ Every 3 months
 ☐ Tele-monitoring
☐ Every 6 months
☐ Other (please specify)

21. How often do you perform the following exams in your patients with this type of Restrictive Chest Disorders adapted to long-term NIV?

	Never	Once	Every month	Every 3 months	Every 6 months	Every year
Full sleep study (Polysomnography)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Portable home sleep study	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Pulsoximetry	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
TcCO ₂	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

10.

* 22. Do you have a formal home care program in your area for patients with Restrictive Chest Disorders on long-term NIV?

- ☐ Scheduled visit at the patient's house by a Nurse
 ☐ Scheduled visit at the patient's house by Home care provider
☐ Scheduled visit at the patient's house by a Respiratory Therapist
 ☐ There is no home care program for Restrictive Chest Disorders patients
☐ Scheduled visit at the patient's house by a Physician
☐ Other (please specify)

23. If you would like to be informed of the results of the survey, please provide your email address below.