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# Long term mechanical ventilation equipment for neuromuscular patients: meeting the expectations of patients and prescribers

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**Running head: Patients' expectations on home mechanical ventilation equipment**

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**ABSTRACT (299 mots)**

**Background**

To maximise the likelihood of successful long term mechanical ventilation (MV) in patients with neuromuscular diseases, ventilators characteristics and settings must be chosen carefully taking into account both medical requisites and the patient's preference and comfort. The general objectives of the survey were 1) to evaluate knowledge and, patient comfort, with their long term MV; 2) to compare patients and prescribers opinions and expectations regarding long term MV; 3) to compare the equipment used by the patients with prescribers present opinion.

**Methods**

Neuromuscular patients receiving long term MV and home MV prescribers in Belgium and France and MV prescribers were asked to respond to a questionnaire survey specifically developed for the study.

**Results**

Completed questionnaires were collected from 209 patients, mean age 35.4±15.9 years (range 3 to 86 years), ventilated since 11 ± 17 year, and 45 MV prescribers. Hundred sixty three (78%) patients correctly designed their MV mode as a volume or a pressure controlled mode and 86% considered their MV as "efficient". When an inspiratory trigger was available, 92% of the patients were able to use it but only 72% were satisfied. Prescribers were more prone than patients to use new technologies, such as an emergency system to release a noninvasive interface (visual analogue scale (VAS/10): 9.2±1.5 vs 6.8±3.3, *P*=0.0001), a humidification system (VAS: 8.6±1.4 vs 7.8±2.6, *P*=0.02), a contactor for providing larger inspiratory volumes (VAS: 8.4±1.7 vs 6.0±3.0, *P*=0.009), an in-built cough assistance mode (VAS: 9.2±1.4 vs 5.5±3.3 *P*=0.00001), new options to improve speech, or new MV modes such as a volume targeted-pressure controlled mode.

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**Conclusions**

Patient's and prescriber's opinion differ about the ideal home ventilator. Patients are less prone to use new technologies, mainly because of a lack of information, underlining the need of regular MV update in patients receiving long term MV.

**Key words:** Home care. Masks. Mouthpieces. Myopathies. Patient-centered care. Respiratory failure.

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## INTRODUCTION

Clinical outcomes of neuromuscular patients using long term mechanical ventilation (MV) depend not only on medical factors, but also on MV equipment as well as the quality of the home support [1, 2]. Selecting the best MV equipment and the optimal settings for each individual patient is crucial for the success of long term MV.

Importantly, although patient comfort is widely recognized as a crucial determinant of MV compliance and efficacy [3], few studies have evaluated the patients' opinion concerning MV equipment. Patient's comfort may be affected by many factors, such as asynchrony between the patient and the ventilator, the inability of the ventilator to cope with the patient's respiratory demand, skin lesions caused by the interface, inappropriate humidification, difficulty to speak during MV, and other causes. Patients receiving long term MV at home are expected to have an "expert" opinion on MV as they have been using MV for years in various daily life conditions. Moreover, the comparison of patient's and prescriber's opinion may be valuable for deciding which technological issues deserve priority. This is of particular interest for neuromuscular patients as this population represents the largest population treated with MV at home [4] and because they are treated either with noninvasive MV or invasive MV via a tracheostomy [1], during night and sometimes daytime [1], which may require different interfaces and ventilator modes.

The general objectives of the present survey were to 1) to evaluate the patients' knowledge about and comfort with their long term MV; 2) to compare patients and prescribers opinions and expectations regarding long term MV; 3) to compare the equipment used by the patients with the prescribers current MV prescription.

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## MATERIAL AND METHODS

### Study questionnaires

Data were collected via questionnaires specifically developed for the study by a scientific committee composed of adult and paediatric teaching-hospital home MV prescribers (from Raymond Poincaré and Trousseau Hospitals); researchers in respiratory physiology, mechanical properties of the respiratory system, and ventilator development and evaluation (from INSERM 955, team 13); MV home care professionals (from ADEP Assistance, a non-profit organization supported by Air Liquide Inc.); MV physiotherapists; representatives of a neuromuscular patient-support organization (*Association Française contre les Myopathies*, AFM); and a representative of ResMed, a company that manufactures ventilators (Saint Priest, France), selected by the scientific board after an industrial application. The patient and prescriber questionnaires were tested prior finalization by 20 representative neuromuscular patients and 5 home MV prescribers in teaching hospitals in the Paris conurbation, respectively. The ability of the patients to understand the questions was checked, as well as the time required to complete the questionnaire which should be less than 90 minutes for both patients and prescribers. The patients questionnaire included patients opinion on long term MV objectives and potential innovations. The prescribers questionnaire was similar to the patients questionnaire and assessed the prescribers opinion on home MV objectives and potential innovations. Special care was taken to formulate similar questions to the prescribers and the patients. The two questionnaires are available on the online repository.

The final patient questionnaires were completed by healthcare professionals between September 2010 and July 2011. When the patient was an adult, the healthcare professional explained all the definitions such as synchronization, effectiveness, comfort of ventilation and others in order to improve the understanding of the questions. When the patient was a

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child, the interview was performed with the parents who assisted the interviewer in order to improve the understanding of the questions.

The study was approved by the French 'Informatique et Libertés' institution (CNIL) in agreement with the French regulation.

**Patient and prescriber recruitment**

The objective was to include at least 200 patients with an equal distribution between patients treated with nocturnal non-invasive MV (NIMV) only, patients treated with NIMV during both night and daytime, and those treated with invasive MV (IMV) via a tracheostomy. Patients were recruited at reference medical centres for neuromuscular diseases in Belgium (Inkendaal) and France (Lille, Grenoble, Marseille, Nice, Paris, and Garches) and at residential facilities for neuromuscular patients in France (Evry, Montpellier, and Angers). The criteria for patient inclusion were 1) a diagnosis of neuromuscular disorder, 2) MV for at least one year, and 3) the willingness and availability to participate to the study (an interview of 90 minutes). The scientific committee decided not to include patients with amyotrophic lateral sclerosis due to the rapid progression of this disease.

The objective was to include at least 40 prescribers. The prescribers were selected at random in the address book of the above-mentioned neuromuscular patient-support organization (AFM). All the 89 AFM-prescribers were contacted by e-mail between July 2010 and December 2010 to assess their willingness to participate to the survey. Forty six prescribers accepted to participate to the survey, one refused, and the others did not respond. The questionnaire was send only in case of acceptance with one prescriber who accepted but did not fill in the questionnaire.

**Statistics**

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Quantitative parameters were described as mean ± standard deviation and as median with the interquartile range (IQR) depending upon the normality of distribution. Qualitative parameters were described as numbers and percentages (%). Responses to items present in both the patient and the prescriber questionnaire, particularly those answered via visual analogue scales (VASs), were compared using the Mann-Whitney test for non-normally distributed variables and the chi-square test or exact Fisher test, as appropriate. Two-sided *p* values smaller than 0.05 were considered significant. All analyses were performed using R software (www.r-project.org/).

## RESULTS

### Patient's and prescriber's characteristics

Completed questionnaires were collected from 209 neuromuscular patients. Only 11 patients refused to answer to the questionnaire. The mean age of the patients was 35.4±15.9 years (range 3 to 86 years) and the mean duration of MV prior the study was 11 ± 17 years. The type of neuromuscular disease and degree of autonomy are reported in Table 1. The ventilators used by the patients are presented in Table 2. NIMV was used during night time (N-NIMV) only by 71 patients and during night and daytime (ND-NIMV) by 71 other patients, including 30 patients who used mouth-piece MV. The remaining 67 (32%) patients had a tracheostomy; among them, 6 patients were not ventilated during daytime.

Completed questionnaires were obtained from 45 prescribers (23 pulmonologists, 12 paediatricians, and 10 intensive care specialists), of which 41 worked in public hospitals. Fifteen prescribers were providers of the patients included in the study. The long term MV experience lasted more than 5 years in 37 prescribers, 5 to 10 years in 11 prescribers, and more than 10 years in 26 prescribers.

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**Knowledge and patient comfort**

Of the 209 patients, 163 (78%) correctly designed their ventilatory mode as volume controlled (VC), pressure controlled (PC), or pressure-regulated volume-control (PRVC) mode, but only 82 (39%) patients knew whether their mode was "assisted", "controlled", or "assist-controlled".

Hundred fifty eight (76%) patients found their MV"comfortable" in term of respiratory sensation (56 (84%) IMV; 56 (79%) ND-NIMV; 46 (65%) N-NIMV) and 22 (11%) patients found their MV "uncomfortable" (of whom 12 N-NIMV, 8 ND-NIMV and 2 IMV) with 29 (14%) patients having no opinion.

Table 3 resumes patients' reports concerning the ability to use the assisted function and its usefulness. The majority of the patients were able to trigger the ventilator and were satisfied by its sensitivity.

Among the 134 patients (73 treated with NIMV and 61 with IMV) who needed daytime ventilation, 73 (54%) patients (including 13 (9%) tracheostomized patients) were able to breathe spontaneously. Of the 60 patients treated with NIMV during daytime and able to breathe spontaneously, 39 (65%) reported feeling better with daytime ventilation, with a decrease in daytime fatigue 37 (62%), an improvement in eating 7 (12%); and/or speech 13 (22%). Of the 13 tracheotomized patients able to breathe spontaneously, 11 (85%) reported feeling better with daytime ventilation, which was reported by the patients to decrease daytime fatigue in 10 (77%) patients, improving eating in 7 (54%) patients, and/or speech in 6 (46%) patients.

**Comparison of patients' and prescribers' opinions**

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With regard to the ventilator performances, except for ventilator efficiency (defined for the patients as the correction of abnormal blood gases [5]) at the expense of comfort, all the ventilator characteristics were quoted as more important by the prescribers than by the patients (Table 4). Allowing the patient to adjust the ventilatory settings was ranked as a low priority by prescribers and patients, but the prescriber's ranking was higher than the patient's ranking. Both patients and prescribers considered that the option of a cough assisted mode was important, but this importance was also ranked higher by the prescribers than by the patients (Table 4).

When the patients were asked if they would accept the implantation of two electromyography electrodes by means of a minor surgical procedure performed under local anaesthesia in order to improve the synchronization of the ventilator to the patient's inspiratory effort, only 24 (11%) patients, including 8 N-NIMV (11%), 7 ND-NIMV (10%), 9 (13%) tracheostomized patients, agreed immediately, 58 (28%), including 16 ND-NIMV, 20 N-NIMV, 22 (11%) tracheostomized patients, answered that they would eventually accept, whereas 112 (54%) patients refused, including 39 (55%) N-NIMV, 42 (59%) ND-NIMV, 31(15%) tracheostomized patients, the remaining patients having no opinion. The prescribers' VAS response concerning the potential interest of this technology (from not useful=0 to very useful=10) was 7.4 ± 2.3.

For secretion clearance, 35 (17%) patients including 7 (10%) N-NIMV, 15 (21%) ND-NIMV, 13 (19%) tracheostomized patients, preferred a mechanical insufflator-exsufflator system; 30 (14%) including 17 (24%) N-NIMV, 9 (13%) ND-NIMV, 4 (6%) tracheostomized patients, an intermittent inspiratory pressure; and 19 (9%) including 3 (4%) N-NIMV, 9 (13%) ND-NIMV, 7 (10%) tracheostomized patients a percussive system, with the remaining 125 (60%) patients having no opinion. Among the prescribers, 30 (66%) prescribers preferred a mechanical insufflator-exsufflator system, 5 (11%) an air-stacking

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method, 4 (9%) an intermittent inspiratory pressure, and 3 (7%) a percussive system, with 2 prescribers having no preference.

Table 4 shows that the prescribers were generally more concerned than the patients about the tolerance of the interface, the importance of a satisfactory headgear, and the possibility to alternate different interfaces in order to change the pressure points. Although prescribers were more concerned than patients for having an appropriate release system allowing the patient to release the interface by him(her)self, 88 (62%) patients among 142 under NIMV, were not able to release the interface by themselves.

Humidification of the circuit was considered more important by the prescribers than by the patients, while the importance of the heating the circuit was ranked similarly by the patients and the prescribers (Table 4).

Concerning mouthpiece ventilation, prescribers were more eager than patients to have a patient-controlled contactor for triggering/stopping insufflation, in order to meet the patient's needs (Table 4).

Among the 67 tracheotomised patients, 5 (7%) used a phonation valve during ventilation. Of the 34 tracheotomised patients able to breathe spontaneously, 15 (44%) patients used a phonation valve, 11 (32%) patients closed their tracheostomy tube completely, and 8 patients kept their tracheotomy tube open during speech.

Figure 1 reports the 67 tracheotomised patients' and prescribers' opinions about technical changes aiming at improving speech. Prescribers were more eager than patients to develop technical innovations to improve speech for these patients.

**Evaluation of the patients 'equipment versus the current prescribers' choice**

One hundred twenty two (59%) of the patients were treated with a VC mode, 82 (39%) patients with a PC mode, and 5 (2%) patients with a PRVC mode. The number of

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patients receiving a VC mode increased in those requiring daily MV (Figure 2). Figure 3 shows the MV modes used by the patients (VC, PC, or a PRVC mode) vs. the prescribers' current MV prescription according to the patient's diagnosis and the MV interface.

Among the 71 patients using NIMV during daytime, 30 used a mouthpiece. The percentage of mouthpiece users during daytime increased with the daily duration of ventilation, from no patient using mouthpiece ventilation in case of a NIMV duration <8 hours/day to 90% of the patients when the duration of NIMV exceeded 20 hours/day (Figure 4).

Table 5 presents the humidification system used by the patients and prescribers' humidification choice. Heated humidifier (HH) was the most used and chosen during NIMV, however heat and moisture exchanger (HME) was the most common humidification system used during IMV, whereas prescription of filter and heated humidification was comparable in this group.

## DISCUSSION

This study is the first to evaluate and compare the prescribers' and patients' expectations regarding long term MV equipment. Interestingly, the opinion and expectations of the patients and the prescribers differ in many aspects. This survey compares also the current MV use in a large group of patients with neuromuscular disease with the prescribers' current MV prescription. Because the technology of the ventilators, circuits, and interfaces change continuously, important differences were observed, due to the fact that the patients' MV equipment may reflects a prescription made several years ago.

### Knowledge and patient comfort

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Only 78% of the patients were able to correctly define their MV modes as VC, PC, or as a combined mode and less than of half of the patients were able to define if their MV mode was assisted, controlled, or both. This shows that most patients have a poor knowledge of their MV and of patient-ventilator synchronization. Interestingly, 92% of the patients who had the possibility to trigger the ventilator insufflations considered that they were able to do it, including all the tracheostomized patients using an assist mode. However 8% of non-tracheostomized patients and 18% of the tracheostomised patients considered their ventilator trigger insufficiently sensitive. On the other hand, the ventilator trigger was reported as "too sensitive" for 5% and 3% of the nontracheostomized and tracheostomized patients, respectively. These results reflect the difficulty to find a trade-off between a trigger which is insufficiently sensitive and avoids the risk of auto-triggering and a trigger which is sufficiently sensitive with the risk of auto-triggering. This aspect is particularly difficult in a population in whom the inspiratory muscle strength may decrease progressively which should lead to a progressive increase in the sensitivity of the inspiratory trigger. An alternative could be the use a new trigger technology such as the detection of the inspiratory effort by means of inspiratory muscle electromyography such as provided with neutrally adjusted ventilator assistance (NAVA) [6]. For a home ventilator, such a trigger should require an implantable device for monitoring inspiratory muscle electromyography. When we asked the patients if they would accept a minor surgical procedure in order to improve their synchronisation with their ventilator, the majority of the patients refused and only 11% of the patients were immediately favourable. Similarly, the majority of the prescribers were not enthusiastic with regard to this possible breakthrough.

**Comparison of patients' and prescribers' opinions**

Both patients and prescribers attached considerable importance to patient/ventilator synchronisation, patient respiratory comfort during MV, and the possibility to have a patient-

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controlled cough support integrated within the ventilator. However, the demand for these characteristics and options was generally stronger for the prescribers than for the patients.

Cough support by air-stacking, which does not require any modification of volumetric ventilators, and intermittent inspiratory pressure may be easily integrated into conventional home ventilators. In contrast, percussive and cough-assist methods will require an additional technology that may translate into increased ventilator weight and size. A review of the literature [7] suggests that the mechanical insufflations-exsufflation technique is able to increase the inspired volume at least as well as the other inflation methods, and peak cough flow is as fast as or faster than manual assisted cough associated to an inflation method, and patients seem to prefer mechanical insufflations-exsufflation method to the other cough assisted methods. Finally, the insufflations-exsufflation method seems to be at least among the best methods for improving airway clearance. A cheap alternative technique could be to use the ventilator to increase the inspired volume associated with manual assisted cough, if ventilator manufacturers are prone to integrate an intermittent inspiratory pressure mode into their ventilator. Accordingly, the possibility to have a patient-controlled cough support integrated within the ventilator was considered important for both patients and prescribers, but again, the prescriber's ranking was higher than the patient's ranking.

The majority of our patients had no preference for any of the different cough assisted methods, which may be explained by a lack of knowledge and experience. Curiously, among the patients who preferred a technique, they preferred the insufflation-exsufflation method followed by the intermittent inspiratory pressure method and the percussive method, whereas an intermittent inspiratory pressure device was the most frequent cough support device used by the patients, followed by the insufflation-exsufflation and the percussive device. This paradox may be explained by the influence of the prescribers of whom 2/3 currently preferred the insufflation-exsufflation method in agreement with the literature data [7].

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Therefore it may be expected that the number of patients using these devices will increase in the near future.

**Evaluation of the patients 'equipment versus the current prescribers' choice**

The choice of the MV mode according to the daily duration of MV is one of the most striking observations of this survey. Indeed, a PC mode, mostly delivered by a bi-level pressure device or a pressure support device, was preferentially used in patients needing only nocturnal MV because these modes are more comfortable than VC modes during non-leakage conditions [8]. Most clinicians prescribe PC modes during sleep, as they allow adjustments of inspiratory time, leak compensation, and improved patient comfort. However, a VC mode predominated in the patients who needed also daytime MV, mainly with a mouthpiece. Although PC mode may be used with a mouthpiece, VC ventilators are more performant for mouthpiece ventilation [9]. Furthermore, mouthpiece ventilation allows the patient to disconnect him(her)self from the ventilator, ideally during expiration, for example to allow speech after each insufflation. PC ventilators, in contrast, respond to disconnection by reviving the turbine in order to maintain the targeted positive pressure level. In addition, by definition, PC modes are not able to generate the high pressures necessary for effective breath-stacking or lung volume recruitment [9, 10]. In contrast, VC ventilators are able to provide both the tidal volume for sustained ventilation and the pressure and volume required for effective lung-volume recruitment and achievement of a maximal insufflation capacity [9, 10]. Nevertheless, another option for improving cough could be to use an independent cough assist device. A second option could be to integrate an additional intermittent-positive pressure mode (see below) into the ventilator.

When we compared the MV modes used by patients and the current prescribers prescription, we observed that the PRVC mode was gaining popularity over the VC mode.

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PRVC ventilation is a new mode that combines a VC and a PC ventilation by using an algorithm that estimates the patient's tidal volume over several breaths, enabling the calculation of the pressure change necessary to achieve the target tidal volume. In addition, PRVC devices are designed to adapt the amount of pressure to changes in respiratory impedance. However, theoretically, PRVC ventilation is not able to cope with unintentional leaks, especially when these leaks vary over time [11], as with NIV via a mouthpiece or ventilation via an uncuffed tracheostomy tube. When unintentional leaks occur during PC ventilation, the inspiratory flow delivered by the ventilator increases to reach the target inspiratory airway pressure whereas the tidal volume decreases [12]. Carlucci et al. [13] have recently observed that, depending on the circuit configuration, many MV devices recognize the delivered volume as the tidal volume. Therefore the inspiratory pressure adjustment may paradoxically decrease to a lower level during unintentional leaks, which worsens the fall in tidal volume. In contrast, when the tidal volume is estimated by the expired volume, the tidal volume can be overestimated [14, 15]. Finally, the majority of the PRVC ventilation devices are currently not reliable during unintentional leaks, underlining the need to check these modes during unintentional leaks.

The fitting and comfort of non-invasive interfaces are crucial for the effectiveness of NIV during sleep [16]. All different types of interfaces were used by the patients. However, despite their advantage of a minimal facial contact which widens the field of vision and allows the use of eyeglasses, few patients used nasal pillows [17]. Until recently, all nasal pillows available in France had intentional leaks and therefore can be used only with bi-level devices, given that any modification of a medical device (e.g., sealing a hole) is prohibited by the French law. Manufacturers are thus encouraged to increase the development of non-leak pillows. Such nasal pillows may be an alternative to mouthpiece MV during daytime [18].

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Another solution to reduce the risk of skin damage is to alternate various types of interfaces. If most prescribers are in favour of this solution, it is not the case for the patients.

As reported in the literature [17], an artificial nose was the most commonly humidification system used by tracheostomised patients, whereas a heated humidification was the most commonly humidification system used by NIMV patients. Humidification and warming of the inspired gas may be used to prevent the adverse effects of cool, dry gases on the airway epithelium. The major cause of humidity loss is an unidirectional flow from the ventilator circuit to the patient because of mouth leaks during nasal NIMV and between the tracheal tube and the trachea when using an uncuffed tube during IMV. These leakages are extremely common in neuromuscular patients [19, 20]. Therefore, although a HME and a HH showed similar tolerance and side-effects during long term MV [21], we prefer a HH because of its greater efficacy than a HME in case of unintentional leaks [22-24]. In addition, as compared to a HH, an HME increases the resistance of the circuit and the dead space and therefore the work of breathing during IMV [25, 26]. However, tracheostomised patient using the wheelchair represents an exception. In this situation, a water tank humidifier built into the circuit and located behind the patient and often above the tracheostomy tube exposes the patient to a risk of water inhalation. A HME, in contrast, is safe.

In conclusion, this large survey underlines the differences between patients and prescribers' concerning the MV equipment and potential improvements. Prescribers are more enthusiasts and more prone than patients to prescribe new technologies such as emergency systems to release the non-invasive interface, contactor for triggering/stopping insufflations or for providing larger inspiratory volume, cough assistance devices, or new technological solutions which could improve speech [27]. For some of these new technologies, such as the PRVC mode, objective clinical benefits are lacking. Patients are more reluctant

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than prescribers to new technologies, and many of them had no opinion which may be explained by lack of information underlining the need of regular MV update in patients receiving long term MV.

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**Figure Legends**

**Figure 1:** Proportion of patients and prescribers prepared to use currently available solutions for improving speech in tracheotomised patients.

**Figure 2:** Distribution of ventilatory mode according to the condition of mechanical ventilation : patients treated with nocturnal non-invasive mechanical ventilation (N-NIMV), patients under NIMV during both night time and daytime (ND-NIMV), and those treated with invasive mechanical ventilation (IMV) via tracheostomy.

**Figure 3:** Modes used by the patients (volume controlled (VC), pressure controlled (PC) and pressure-regulated volume-control (PRVC) vs. prescriber opinions in three diagnostic groups, according to ventilation interface (only the most frequent conditions were taken in account): Duchenne muscular dystrophy patients with nocturnal non-invasive mechanical ventilation (NIMV), nocturnal and daytime NIMV, or nocturnal and daytime invasive mchanical ventilation (IMV); myotonic dystrophy patients with nocturnal NIV; and spinal muscular atrophy patients with IMV.

**Figure 4:** The percentage of mouthpiece users and other interface users during daytime according to patient's ventilatory dependance.

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**Table 1:** Patient characteristics and degree of autonomy

| General characteristics | Number of patients | % |
|---|---|---|
| Females | 69 | 33 |
| Children (≤16 years) | 24 | 11 |
| Institutionalized | 40 | 19 |
| **Type of neuromuscular disease** | | |
| Duchenne and Becker muscular dystrophies | 71 | 35.7 |
| Myotonic dystrophies | 35 | 17.6 |
| Spinal muscular atrophies | 20 | 10.1 |
| Congenital myopathies | 15 | 7.5 |
| Sarcoglycanopathies | 8 | 4.0 |
| Proximal myopathies | 6 | 3.0 |
| Other myopathies | 8 | 4.0 |
| Other neuropathies | 30 | 15.1 |
| **Degree of autonomy** | | |
| Able to wash him(her)self without help | 47 | 22.5 |
| Needs help for washing | 55 | 26.3 |
| Washed by another person | 107 | 51.2 |
| Able to eat without help | 73 | 35.6 |
| Needs help for eating | 113 | 55.1 |
| Fed by a tube | 19 | 9.3 |
| Able to stand without help | 43 | 21.3 |
| Able to stand with help | 115 | 56.9 |
| Unable to stand | 44 | 21.8 |
| Able to walk without help | 39 | 19.8 |
| Able to walk with help | 27 | 13.7 |
| Cannot walk | 143 | 68.4 |
| Uses a manual wheelchair | 48 | 24.4 |
| Able to drive a car | 19 | 9.5 |
| Drives a power wheelchair | 145 | 72.5 |
| No mobility without help | 36 | 18 |

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**Table 2. Ventilators used by the patients**

| Ventilator type | Number of patients |
|---|---|
| EOLE 3 (ResMed SA, Saint Priest, France) | 67 |
| ELISEE 150 (ResMed SA, Saint Priest, France) | 32 |
| VIPAP III (ResMed SA, Saint Priest, France) | 6 |
| VIPAP IV (ResMed SA, Saint Priest, France) | 14 |
| VS Integra (ResMed SA, Saint Priest, France) | 1 |
| VS ultra (ResMed SA, Saint Priest, France) | 8 |
| LEGENDAIR (COVIDIEN Courtaboeuf, France) | 28 |
| AIROX SMART AIR (COVIDIEN Courtaboeuf, France) | 4 |
| AIROX Home 2 (COVIDIEN Courtaboeuf, France) | 1 |
| PB560 (COVIDIEN Courtaboeuf, France) | 3 |
| VIVO 40 (Breas Medical, Saint Priest, France) | 8 |
| VIVO 50 (Breas Medical, Saint Priest, France) | 1 |
| TRILOGY 100 (Respironics France, Carquefou, France) | 9 |
| BiPAP Synchrony (Respironics France, Carquefou, France) | 26 |
| BiPAP Harmony (Respironics France, Carquefou, France) | 1 |

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**Table 3:** Patients'reports concerning the inspiratory trigger

| **Assisted mode available** | **N-NIMV (n = 60)** | **ND-NIMV (n= 56)** | **IMV (n = 38)** |
|---|---|---|---|
| Ability to use the assisted mode | 58 | 50 | 33 |
| The inspiratory trigger is (too /satisfactory/insufficiently) sensitive | 3/41/6 | 2/39/3 | 1/27/6 |
| **When do you decide to increase the respiratory frequency during mechanical ventilation ?** | | | |
| when seeking for more air | 19 | 16 | 14 |
| during daily activity like eating or having bowel movement | 3 | 7 | 7 |
| for speaking | 4 | 5 | 4 |

Abbreviations : N-NIMV : nocturnal non-invasive mechanical ventilation, ND-NIMV : NIMV during both night time and daytime, IMV invasive mechanical ventilation via tracheostomy

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**Table 4:** Visual analogue scale scores (from not important=0 to very important=10) assigned by patients and prescribers regarding ventilator performance, non-invasive interface, humidification and heating and mouthpiece condition.

| **Questions** | **Patients** | **Prescribers** | |
|---|---|---|---|
| **Ventilator performance** | **Mean±SD** | **Mean±SD** | ***p* value** |
| Patient/ventilator synchronisation | 8.3±1.8 | 9.0±1.6 | 0.01 |
| Greater effectiveness at the expense of decreased comfort | 7.9±2.0 | 6.5±2.2 | 0.0004 |
| Greater comfort at the expense of decreased effectiveness | 7.5±2.3 | 7.9±1.9 | NS |
| Patient able to adjust the settings according to needs | 3.6±3.7* | 5.5±2.8 | 0.0003 |
| Cough support system built into the ventilator | 5.5±3.5 | 9.2±1.4 | 0.00001 |
| Cough support system built into the ventilator and under patient control | 8.0±2.8 | 9.3±1.2 | 0.00001 |
| Contactor for providing larger inspiratory volumes for coughing | 6.0 ± 3.0 | 8.4 ± 1.7 | 0.009 |
| **Non-invasive interfaces state Number of patients = 142** | **Mean±SD** | **Mean±SD** | **P** |
| Good tolerance of the interface | 9.3 ± 1.1 | 9.8 ± 0.5 | 0.0001 |
| Satisfactory system for holding the interface in place | 8.9 ± 1.6 | 9.6 ± 0.7 | 0.0001 |
| Two different interfaces to alternate pressure points | 6.8 ± 3.5 | 9.2 ± 1.1 | 0.0001 |
| Ability to independently secure the interface | 8.2 ± 2.5 | 8.2 ± 1.9 | NS |
| Ability to independently remove the interface | 8.8 ± 1.9 | 9.0 ± 1.3 | NS |
| Emergency release system built into the interface | 6.8 ± 3.3 | 9.2 ± 1.5 | 0.0001 |
| **Humidification and heating** | **Mean±SD** | **Mean±SD** | **P** |
| Humidification system | 7.8±2.6 | 8.6±1.4 | 0.02 |
| Heating of the ventilator circuit | 6.7±3.0 | 6.9±2.3 | NS |
| **Mouthpiece condition state Number of patients = 30** | **Mean±SD** | **Mean±SD** | **p value** |
| Contactor for triggering/stopping insufflation | 4.2 ± 4.1 | 7.1 ± 2.4 | 0.02 |
| Off contactor for extending battery life | 7.0 ± 4.1 | 7.7 ± 2.2 | NS |

*When comparing the 3 groups of patients no significant difference was observed, except for the "Patient able to adjust the settings according to needs" which was higher in tracheotomised patients than in N-NIMV (VASS : 4.4±3.7 versus 2.7±3.3); P =0.008)

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**Table 5:** Patients' humidification using and prescribers' humidification choice

| **Humidification systems used by the patients** | **N-NIMV** | **ND-NIMV** | **IMV** |
|---|---|---|---|
| | **n = 71** | **n = 71** | **n = 67** |
| Heat and moisture exchanger | 9 | 7 | 34 |
| Non-heated humidifiers | 11 | 8 | 6 |
| Heated humidifiers | 21 | 16 | 20 |
| No humidification system | 30 | 40 | 7 |
| **Humidification systems proposed by the prescribers** | | **n= 45** | |
| | **NIMV** | | **IMV** |
| Heat and moisture exchanger | 16 | | 27 |
| Non-heated humidifiers | 4 | | 5 |
| Heated humidifiers | 38 | | 32 |
| No humidification system | 4 | | 0 |

Multiple choices were possible for the prescribers
Abbreviations : see Table 3

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Figure 1 R2

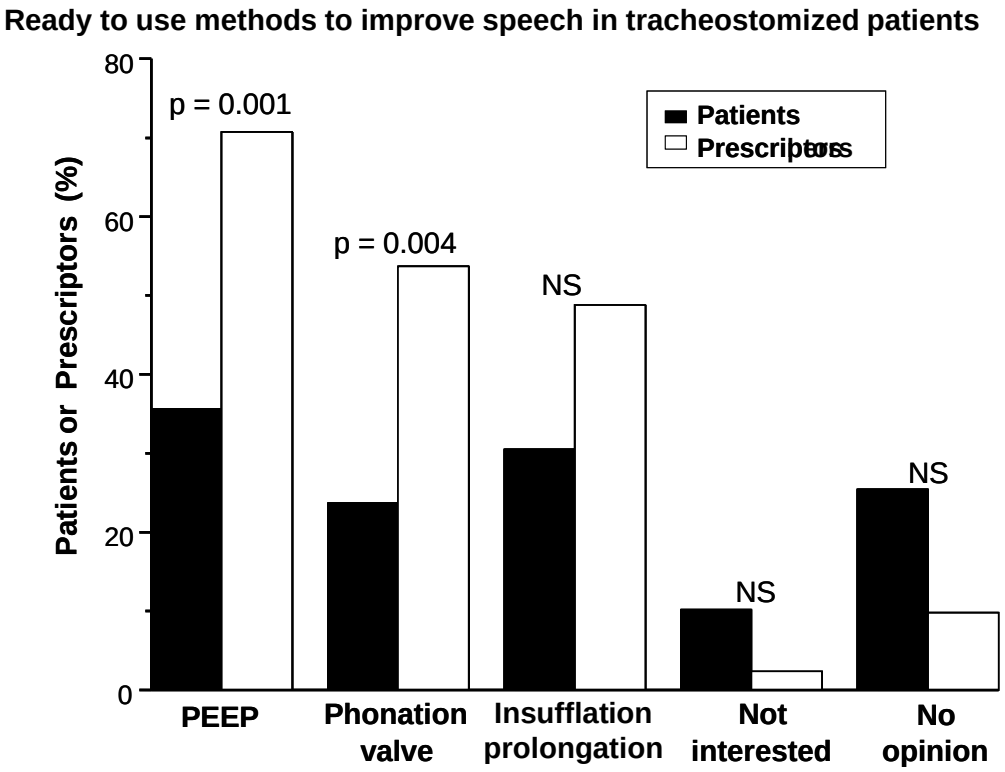

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Figure 2 R3

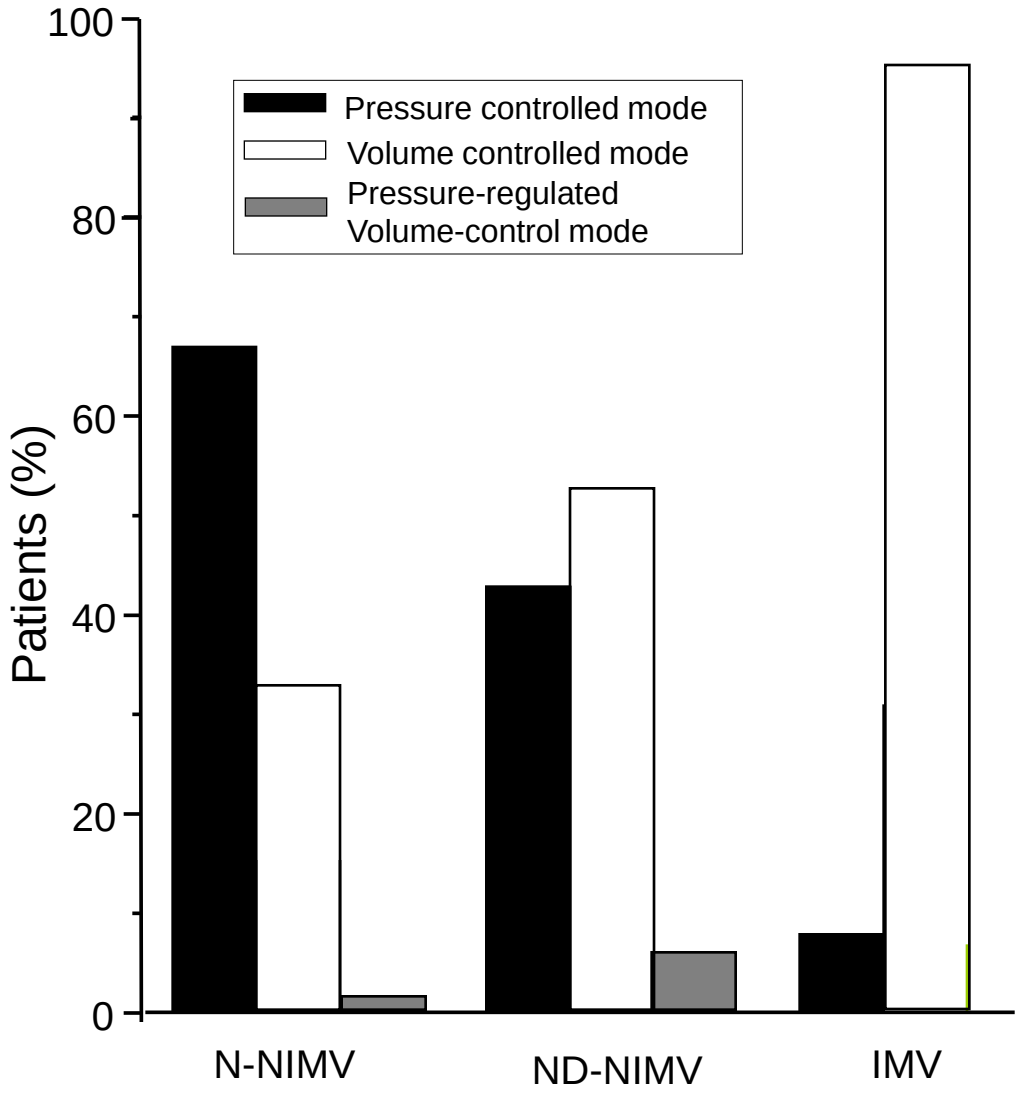

Figure 3 R3

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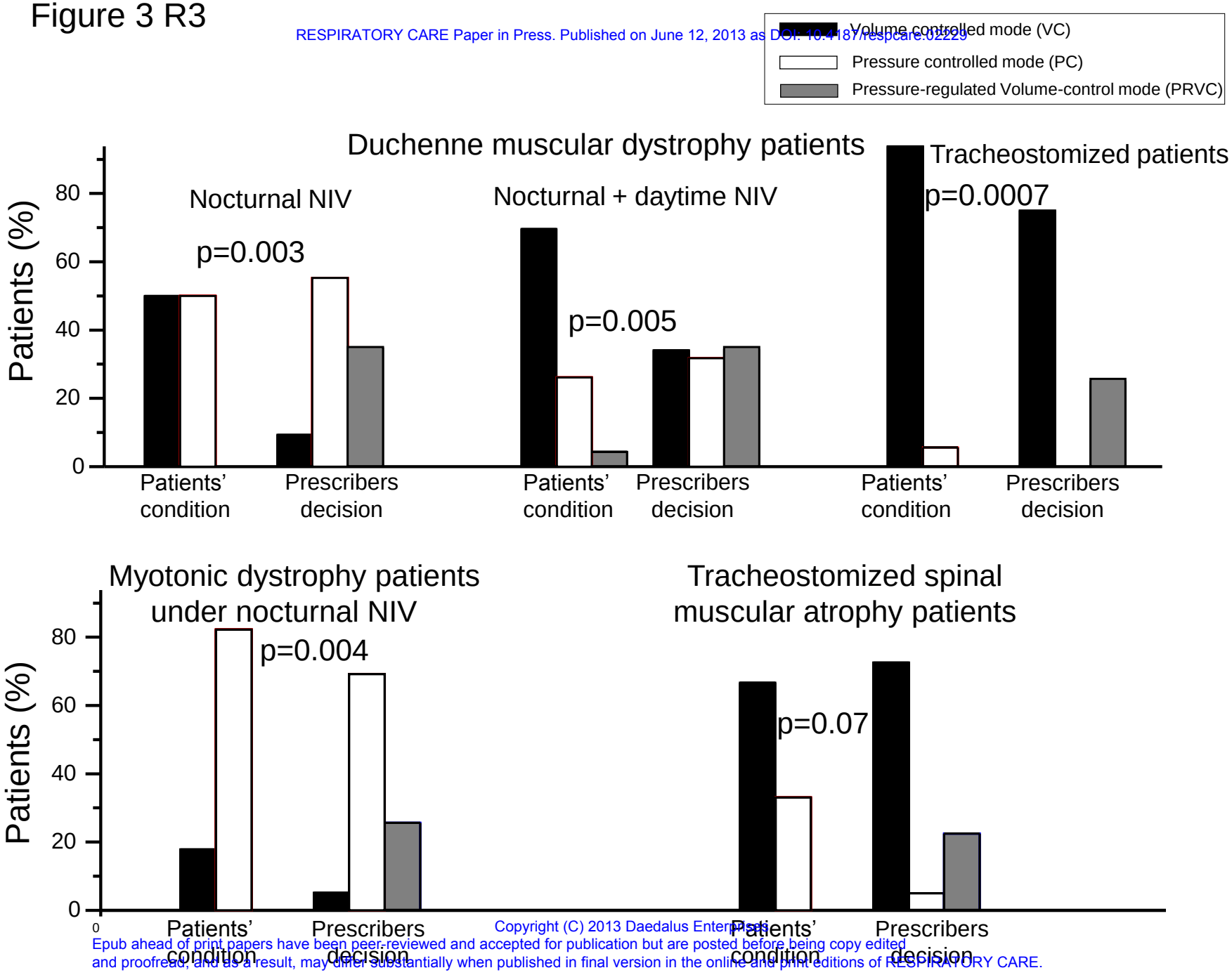

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Figure 4 R2