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To cite this article: Carla Ribeiro, Rita Gomes, Alexandra Carreiro, Ana Luísa Vieira, Bárbara Seabra, Bebiana Conde, Carla Nogueira, Cristina Jácome, Joana Lages, Margarida Aguiar, Mónica Grafino, Paula Pamplona, Ana Cysneiros, Célia Durães, Cidália Rodrigues, Cláudia Pimenta, Cristina Cristóvão, Daniela Rodrigues, Diva Ferreira, Filipe Gonçalves, Helena Ramos, João Cravo, João Paulo Silva, Karl Cunha, Lucía Méndez, Mafalda Van Zeller, Márcia Araújo, Margarida Barata, Margarida Raposo, Margarida Redondo, Maria Jacob, Maria João Araújo, Miguel R. Gonçalves, Miguel Guia, Nuno Faria, Pedro Viegas, Sara Conde, Marta Drummond & Paula Pinto (2025) Home mechanical ventilation in adults: Clinical practice recommendations from the Portuguese Respiratory Society home mechanical ventilation assembly, *Pulmonology*, 31:1, 2598915, DOI: [10.1080/25310429.2025.2598915](https://doi.org/10.1080/25310429.2025.2598915)

To link to this article: <https://doi.org/10.1080/25310429.2025.2598915>



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Home mechanical ventilation in adults: Clinical practice recommendations from the Portuguese Respiratory Society home mechanical ventilation assembly

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ABSTRACT

Background: The use of home mechanical ventilation (HMV) has been increasing worldwide, driven by widening of clinical applications and improved patient survival rates. In Portugal, recent data indicate an even faster growth, although national HMV recommendations have remained unchanged for 25 years.

Research question: We aimed to provide an update in clinical practice guidelines for HMV in adults, applicable to the Portuguese context, grounded on the latest available evidence and experts' opinion.

ARTICLE HISTORY

Received 8 December 2024
Accepted 16 November 2025

KEYWORDS

Home mechanical ventilation; recommendations; clinical practice; expert consensus

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 Supplemental data for this article can be accessed online at <https://doi.org/10.1080/25310429.2025.2598915>.

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Study design and methods: A multidisciplinary panel with experience in HMV in the adult population was assembled. A comprehensive literature search was conducted during March 2023 regarding specific topics: equipment, ventilatory modes and interfaces, HMV initiation, follow-up and monitoring, disease specificities (neuromuscular diseases, obesity-hypoventilation syndrome, restrictive chest wall diseases; chronic obstructive pulmonary disease, and other diseases), home mechanical invasive ventilation, and palliative and end of life care. A 2-round Delphi process was conducted to establish consensus among panel members. A minimum agreement threshold of 80% was required.

Results: Out of 88 recommendations initially included in the Delphi process, 61 were selected by consensus.

Conclusion: Final recommendations grounded in the current level of evidence are outlined, and the key limitations and proposals for future research are discussed.

Introduction

Home mechanical ventilation (HMV) is indicated for patients with severe chronic hypercapnic respiratory failure from various causes aiming to both improve survival and symptom burden.^{1,2}

Worldwide, the use of HMV has been increasing due to expanded indications, improved patient survival rates and enhanced healthcare organisation.^{3–5} In the Eurovent survey conducted in 2001, Portugal had an estimated 933 ventilated patients (9.3 per 100 000 inhabitants).⁶ By December 2020, the number of patients with HMV had risen to 13 448, resulting in an estimated prevalence of 130.5 per 100 000 inhabitants.⁷ This growth in Portuguese prevalence is greater than described in other countries.³

Initially, the predominant indications for HMV were neuromuscular diseases (NMD) and restrictive chest wall disorders (RCWD). However, there has been a significant increase in HMV in patients with obesity hypoventilation syndrome (OHS) and chronic obstructive pulmonary disease (COPD).^{3,4,8} In addition, there has also been substantial contribution of research and technological advances in the HMV field, focusing on broader non-invasive monitoring, new ventilatory modes, improved interfaces, and patient-oriented outcomes.

Despite these changes, recommendations for HMV in the adult population in Portugal have remained unchanged over the last 25 years.^{9,10} Updated recommendations from international societies such as the European Respiratory Society, German Respiratory Society and Swiss Society of Pulmonology are being used to inform clinical practice.^{11–13} However, some of the recommendations do not align with the reality of Portuguese healthcare organisation and reimbursement policies.

In Portugal, home respiratory care such as oxygen and positive airway pressure therapy (continuous, bilevel or servoventilation) are totally supported by the National Health Service and provided through home respiratory care providers that are contracted to provide equipment, maintenance as well as a specified minimal number of home visits and periodic monitoring services.¹⁴ For other therapies or strategies not specified in the document, reimbursement is currently not available, and physicians may be less likely to prescribe them, despite potential clinical benefits. While adapting to this changing reality, practice patterns have evolved over time, resulting in high heterogeneity in prescription patterns, both between countries and within the same country.^{3,4,6,15,16}

There is a need for updated national recommendations to inform prescription practices based on high-quality evidence. In areas where evidence is lacking, it is vital to gather expert opinion informed by country-specificities and regional differences. Incorporating expert insights can help address gaps in knowledge and improve the overall quality of care for patients requiring HMV.

Our aim was to produce updated clinical practice recommendations on HMV for the adult population adapted to the Portuguese Healthcare system, grounded on the latest available evidence and experts' experience and opinion.

Methods

Expert panel

A multidisciplinary panel, co-chaired by C. Ribeiro and R. Gomes, consisted of 38 members with experience in HMV, including 31 pulmonologists, 2 dual pulmonologists/intensivists, 2 physiotherapists, 1 nurse, 1 cardiopulmonologist and 1 patient association representative who is also a physiotherapist and nutritionist. Any conflicts of interest were declared prior to the commencement of the work. Initial meetings were used for defining chapters to be addressed: 1. equipment, ventilatory modes and interfaces; 2. HMV initiation; 3. follow up and monitoring; 4. Disease specificities (NMD; OHS; RCWD; COPD; Other diseases (cystic fibrosis, bronchiectasis, interstitial lung disease); 5. home mechanical invasive ventilation; 6. palliative and end of life (EoL) care. The methodologist (C. Jácome) supported the methodological approach to produce the recommendations but did not participate in their formulation. Groups of 3–4 members were formed focusing on specific areas, with members allocated to each group based on their preferences and areas of expertise. A coordinator was selected for each group. The chairs and group coordinators held bi-monthly virtual meetings between January and November 2023. The entire panel met twice virtually and once in person.

Literature searches

A comprehensive literature search was conducted March 2023 PubMed and Cochrane databases. Search results were initially independently screened by title and abstract by at least 2 members of each group for eligibility. In a second phase, selected papers were screened by full text. Relevant systematic reviews and guidelines were also consulted for additional references. The search process and detailed search strategies are summarised in supplementary material.

Evidence synthesis and formulating recommendations

Each group outlined the relevant evidence related to their area first in a summary table. Then a summary text and up to 10 recommendations were drafted. Although assessment of the quality of evidence was not conducted, recommendations backed by a substantial number of high-quality studies were articulated using phrases such as ‘We recommend’ or ‘should’. In contrast, those supported by fewer studies or based on expert opinion were expressed with terms like ‘We suggest’, ‘could’, or ‘may’.

Delphi process

A 2-round Delphi process was conducted to establish consensus among the 38 panel members. In the first and second rounds, each panel member rated the initial 88 recommendations on a scale from 1 (completely disagree) to 5 (completely agree). An option of ‘non-applicable’ was provided, to allow for the possibility of participants providing opinions within the realm of their expertise. Participants could provide suggestions for improving recommendation statements. For the second round, an international panel of experts composed of 8 pulmonologists and 1 respiratory physiotherapist, all involved in research and members of previous guidelines, were invited to participate to gain broader insight. A minimum agreement threshold of 80% of respondents rating the recommendations with a 4 or 5 was required for acceptance. Out of 88, 61 obtained the consensus required and were included.

Results

This section is organised according to the 6 pre-established topics. For each specific topic, a summary text and a table with the recommendations achieving consensus are provided.

Equipment, ventilatory modes and interfaces

Equipment

Equipment requirements depend on patients' disease needs and must adapt to different life settings. When choosing a ventilator, it is crucial to consider patients who require assistance to manage ventilator interruption, such as a power failure or equipment malfunction.¹⁷ This is especially important for those who are ventilator-dependent. Additionally, patient mobility and geographic location are important factors that further justify the need for an external battery, as the latter contributes to the overall effectiveness and safety.³ For those requiring exclusive nocturnal ventilation, an internal battery is not mandatory, unless they live in areas with inconsistent energy supply.³ For those requiring between 8 and 16 hours of ventilation per day a ventilator with internal batteries is suggested.³ Two life-support ventilators are recommended for patients requiring over 16 hours of ventilation daily, equipped with long-lasting internal batteries, and alarm settings to alert caregivers of any potential issues.^{17,18} Life-support ventilators should enable patient mobility and allow the configuration of different settings and programs, allowing for straightforward switching between ventilatory modes. Equipment with advanced options such the visualisation of flow-time and pressure-time waveforms and settings including inspiratory time, rise time, trigger, and cycle may be necessary to ensure comfort and patient-ventilator synchrony.¹⁷⁻¹⁹ An external battery may be necessary, regardless of the duration of daily ventilation, particularly in certain diseases, such as amyotrophic lateral sclerosis (ALS) or cervical spinal cord injury, given their potential dependence on HMV.

Poor equipment hygiene may expose patients to an increased risk of respiratory tract colonisation with potential for respiratory infections. Appropriate handling should be performed between patients (in reusable devices) and regular cleaning reinforced regularly with patients.²⁰⁻²²

Ventilatory modes

Research has demonstrated the effectiveness of different ventilatory modes in chronic respiratory conditions, improving gas exchange, reducing hypercapnia, and enhancing health-related quality of life (HRQoL).²³⁻²⁷ Given the variability in algorithms and potential discrepancies in the nomenclature of ventilatory modes across different ventilator manufacturers, meticulous attention must be paid to ensure the chosen parameters and settings are compatible with the patient's respiratory mechanics and underlying pathology.²⁸⁻³¹ These modes should be tailored to the patient's needs, with options for night-time and daytime use, exercise, and mouthpiece ventilation, if necessary.^{32,33} Real-life studies show that pressure support with backup respiratory rate or spontaneous-timed mode is the most used for HMV.^{1,4} A survey on HMV users in four European countries found bilevel ventilators to be the most preferred due to their smaller size, lower cost, and ease of use.³⁴ However, there is insufficient evidence to demonstrate the superiority of hybrid or volume modes over spontaneous-timed mode.^{23-27,35,36} Therefore, pressure mode is considered a suitable starting point for initiating nocturnal ventilation, setting hybrid or volume modes aside for patients who do not tolerate or respond inadequately to pressure mode.^{4,17,34}

Challenges arise when selecting the mode for programming mouthpiece ventilation (MPV). In this form of ventilation, the patient triggers breaths through a dedicated mouthpiece, making it ideal for those who can initiate breaths but require ongoing ventilatory support, particularly in cases of NMD.^{32,33,37} It improves comfort and autonomy compared to other methods,³⁷⁻³⁹ but demands patient cooperation and may not be suitable for all patients.^{32,33,37} Challenges include irregular breathing patterns, an open circuit and the need for mouth closure.^{33,37} Using volume ventilation, MPV can facilitate air-stacking and optimise airway clearance techniques (ACT). Additionally, MPV may support speech production and improve coordination between breathing and swallowing, which are particularly relevant in patients with NMD.^{32,37,40} On the other hand, in pressure ventilation excessive leaks may delay transition to expiration as the flow rate is sustained to reach the defined pressure, not allowing air stacking.^{33,37,41} Some manufacturers already offer a dedicated mode, that automatically silences alarms and improves trigger sensitivity, delivering on-demand airflow according to the patient's preferences.

Interfaces, circuits and accessories

For HMV success, the right interface must be chosen.^{42,43} The mask is a key factor for patient comfort and long-term compliance with HMV. There is no perfect interface, and its choice requires careful assessment of

patient characteristics and ventilation modes. Efforts should be made to minimise air leakage, maximise patient comfort, and optimise patient-ventilator interaction.

Breathing circuit characteristics may differ between ventilators, and the choice of circuit should be adapted to the patient's configuration and the ventilator's technical specifications. In HMV, three main circuit types are commonly used: (1) single-limb with intentional leak (used with passive exhalation ports and common in non-invasive ventilation), (2) single-limb with exhalation valve (allowing better control of expiratory flow), and (3) dual-limb circuits (more frequently used in life-support ventilators and invasive settings). Each type has implications for monitoring accuracy, humidification setup, and leak compensation.¹⁷ Healthcare professionals should be aware the impact mask design has on dead space and on patient's inspiratory effort during HMV.^{44,45}

The amount of oxygen supplied under HMV is determined by several factors including oxygen flow rate, location of oxygen entrainment, ventilator pressures, flow rates, interface leakage and the location of the exhalation port.¹⁹ If, for clinical reasons, the amount of inspired oxygen must be raised quickly, in addition to raising the oxygen flow rate, the options include optimising the location of oxygen entrainment or adjusting expiratory positive airway pressure (EPAP), usually maintaining pressure support. Supplemental oxygen is delivered more effectively through the mask than at other points in the ventilator circuit, but this is usually reserved for the acute setting.⁴⁶ At home, the panel considers oxygen supplementation should preferably be applied near or into the ventilator due to lower risk of leaks caused by circuit disconnection.

Some patients experience significant dehydration of the airway mucosa, increasing nasal resistance, which can hinder compliance with ventilatory therapy. Humidification may improve adherence.^{47–49} Heating and humidification may be necessary to avoid the adverse effects of cold and dry gas, and some authors favour heated humidification over moisture exchangers due to the decrease in dead space.⁴² However, when a humidifier is used, careful monitoring should be carried out for possible pressure drops leading to insufficient assistance.⁵⁰

Recommendations are presented in Table 1.

Initiation

Assessment

The aim of HMV is, whenever possible, to correct nocturnal hypoventilation (attaining normocapnia) and respiratory sleep disturbances.^{11,30} Proper assessment of respiratory failure and comorbidities is crucial for successful titration. Therefore, a detailed medical history and physical examination should be conducted, looking for signs of chronic respiratory failure (CRF), such as cyanosis, tachypnoea, tachycardia, carbon dioxide (CO₂)-associated vasodilation, and erythrocytosis.¹² The main diagnostic tests used are pulmonary function tests (PFT), sleep studies, daytime arterial blood gas (ABG) analysis

Table 1. Recommendations for equipment, ventilatory modes and interfaces.

1.I	We recommend standard non-battery ventilators for patients who only require ventilatory support at night. For those needing more than 16 hours of support per day, life support ventilators with a built-in battery should be prescribed, along with a second backup ventilator and registration as priority clients with the electrical company. Patients requiring ventilation for 8 to 16 hours per day may benefit from a ventilator equipped with a built-in battery.
1.II	When initiating mouthpiece ventilation, we suggest the use of volume mode as it allows intermittent ventilation without attempting leak compensation.
1.III	The choice of interface should be individualized based on patient characteristics to promote ventilation effectiveness and adherence to treatment. We recommend oronasal masks if nocturnal oral breathing predominates, resulting in high leakage, and if nasal obstruction and congestion do not respond to treatment. Nasal masks may be favoured in patients with upper airway mask-induced obstructions, daytime ventilation and excessive mucous production, such as bronchiectasis and cystic fibrosis.
1.IV	To minimize contamination risk, we recommend daily cleaning of the interface and weekly cleaning of the circuit, harness and ventilator with a neutral detergent. If a humidifier is installed, it should be washed daily, and bottled/distilled water should be used in the container.
1.V	When selecting circuits for HMV, we recommend choosing those that are lightweight, resistant to occlusion, flexible, offer no resistance to airflow, with secure connections and minimal dead space.
1.VI	Oxygen supplementation should preferably be applied near or into the ventilator due to lower risk of leaks caused by circuit disconnection.
1.VII	In patients with clinically relevant mucosal dryness, we recommend active humidification with heated humidifiers to improve compliance and comfort.

preferably at room air, nocturnal oximetry and/or transcutaneous capnography (tCO₂). The type of diagnostic sleep study should be tailored to the patients' specific condition and the available resources.

Setting

The place of initiation whether at home, at the hospital ward, sleep laboratory or outpatient clinic should be selected considering local resources, expertise, availability of technical support (such as telemonitoring capacity) and local policies. Until recently, HMV adaptation during an elective inpatient admission or laboratory titration was considered the gold standard, as it allowed for the assessment of overnight physiological responses and treatment titration. However, it required extensive night-time measurements, a high demand for hospital beds and specialised staff, generating delays and increasing costs. As a result, it has not become common practice in many countries, including Portugal.^{15,51} Recent randomised controlled trials (RCTs) have shown that home adaptation with telemedicine is feasible, with similar efficacy but lower costs compared to inpatient adaptation in patients with COPD, NMD and other restrictive disorders.^{52,53} In patients with NMD, studies with outpatient adaptation without telemedicine have also shown satisfactory results with similar adherence, improvements in gas exchange, number of unscheduled visits and side-effects, reduced waiting time to HMV initiation, similar or better HRQoL and symptom control compared to hospital-based set-ups strategies.^{54–60} An uncontrolled Portuguese trial showed that in patients with COPD, outpatient adaptation resulted in adequate compliance, significant improvement in ABG and a positive impact in short-term HRQoL.⁶¹ Another study including all diseases reported a favourable patient perception of outpatient adaptation to HMV with good patient-physician communication and significant health reported gains.⁶² Finally, optimal settings and addressing of side effects may require time (days/weeks) and serial adjustments which is easier in the outpatient than inpatient setting. In the presence of unstable comorbidities (e.g., patients with COPD and severe heart failure or at risk of cardiac dysfunction) and acute or acute-on-chronic respiratory insufficiency, hospitalisation for adaptation is advisable.⁶³

Team

We recommend that HMV be initiated and managed by a multidisciplinary team experienced in HMV. The team's motivation, experience, and commitment are more important than the service location.^{59,60} Treatment goals should always involve patients and caregivers through shared decision-making facilitated by dynamic, effective and sensitive communication.⁶⁴ It is crucial to prioritise the interests of patients and caregivers, who need to understand and be actively involved in the treatment plan.

Monitoring tools

During initiation, monitoring tools are recommended to optimise ventilation parameters. These include oximetry, transcutaneous capnography and real-time monitoring of ventilator data. When available, real-time ventilator data, particularly flow curves, provide immediate feedback on ventilatory parameters and assess patient-ventilator asynchronies (PVA). Routine polysomnography (PSG) to identify PVA is resource- and time-consuming, and there is insufficient clear data on the importance of thorough PVA assessment and management for HMV efficacy and clinical outcomes.^{31,65,66} Therefore, we recommend laboratory PSG titration of HMV be reserved for difficult-to-treat patients such as patients with comorbidities and/or asynchronies that limit ventilation efficacy or adherence and/or hypoventilation that persists despite basic parameterisation. A French group proposed five goals to assess successful HMV initiation: a daily use > 4 h/day, an improvement in gas exchange, HRQoL and sleep quality and an absence of side effects.⁶⁷ However, they found that even in an experienced centre only 68% of patients achieved 3 out of the 5 predefined goals and only 9% achieved all five goals. This led them to conclude that attaining all goals defining successful initiation is rarely achieved in real life.⁶⁷

Education

Patients and caregivers must be thoroughly educated about the proper use and maintenance of the ventilator. This education should cover how to use ventilation equipment, its benefits for preferred outcomes – be it survival, symptom control, HRQoL or other – and practical skills such as maintenance, side-effect minimisation and troubleshooting, preferably through both verbal and written information. Patients

should also be trained how to identify warning signs, such as early symptom exacerbation as well as criteria for increasing ventilation periods or seeking medical help.

Recommendations are presented in Table 2.

Follow up and monitoring

Timing

There is limited data on the benefits of follow-up and monitoring. However, it is widely considered that regular follow-up for HMV is essential to ensure treatment effectiveness, safety, and tolerance, and should be tailored to patient's needs and considering underlying disease progression. Follow-up modalities have been reviewed in detail by several expert groups including clinical evaluation, ventilation assessment, adherence, side effect management, and equipment maintenance.^{31,68–74} Most recommendations regarding follow-up periodicity are based on experts' opinion and surveys.^{12,13,69,75,76} Windisch *et al.* recommended that the first re-evaluation should occur within the first 4–8 weeks of implementing HMV.¹² Repeated evaluations are usually performed every 3–12 months depending on degree of stability and progression of the underlying disease.^{12,13}

Monitoring

Symptom persistence (including dyspnoea, nocturnal hypoventilation symptoms, poor sleep quality), lack of ABG improvement and HMV side effects should prompt treatment adjustments.^{69,77} Simultaneously, visits should be harnessed to review equipment hygiene and functionality as well as patient knowledge on correct ventilator use and maintenance.⁶⁹

Normalising daytime ABG is a major physiological goal for patients under HMV. However, HMV overall efficacy cannot be assessed by this measurement alone. Nocturnal gas exchanges need to be assessed by nocturnal pulse oximetry and/or by transcutaneous capnography.

The SomnoNIV group recommends pulse oximetry (including visual inspection of traces) be used as a screening tool in stable patients without supplemental oxygen to identify and exclude patients who do not require more detailed, expensive and time-consuming investigations.⁷² Additionally, overnight transcutaneous capnography is now considered as a reasonable surrogate for nocturnal paCO_2 by the American Academy of Sleep Medicine.⁷⁸ According to the SomnoNIV group, there are large discrepancies in parameters provided by the different equipment software.⁷² Relevant parameters for monitoring HMV have yet to be clearly defined and recommendations in this field are much needed. Ventilator data useful for HMV adjustments include compliance, leaks, tidal volume, respiratory rate, apnoea-hypopnoea index and PVA. In the absence of equipment parameter validation, information provided should only be considered as an indicator of trends without any guarantee as to linearity of its estimations.⁷²

Table 2. Recommendations for initiation of home mechanical ventilation.

- 2.I In the initial evaluation, we recommend a thorough medical history and physical examination looking for signs and symptoms of hypoventilation. This evaluation should include pulmonary function tests (PFTs), diurnal arterial blood gas (ABG) analysis, sleep study (when appropriate) and nocturnal oximetry or transcutaneous oxycapnography.
- 2.II We suggest undergoing a sleep study (type I, II or III depending on patients' characteristics and local availability) whenever a concomitant sleep-related breathing disorder is suspected.
- 2.III A multidisciplinary team is advised for initiation and follow up comprising healthcare professionals with experience in respiratory monitoring and provision of HMV, namely: pulmonologists experienced in HMV as well as physiotherapists, specialist nurses, cardiopulmonary physiologists and, when appropriate, a palliative care clinician. Treatment and goals of care should always involve patients and caregivers (shared decision-making), especially in patients with neuromuscular diseases and cystic fibrosis.
- 2.IV We suggest choosing the place of initiation and follow-up (home, sleep laboratory or outpatient clinic) considering local resources, expertise, technical support (such as telemonitoring) and local policies.
- 2.V We recommend inpatient initiation in the presence of unstable comorbidities and acute-on-chronic respiratory insufficiency.
- 2.VI In resource-limited settings, we recommend laboratory polysomnographic titration of HMV be reserved for difficult to treat patients (e.g., those with comorbidities and/or asynchronies that limit ventilation efficacy or adherence).
- 2.VII We suggest using oximetry, transcutaneous capnography and, when available, real-time ventilator data monitoring for HMV titration. Oximetry and diurnal ABG analysis should be used in the absence of capnography availability.
- 2.VII We suggest titration of optimal ventilator parameters and settings to achieve ventilatory efficacy including improvement in gas exchange, health-related quality of life (HRQoL), symptoms and patient comfort. If maladaptation or hypoventilation persists after optimization of ventilatory parameters, laboratory polysomnography may be required to assess and correct asynchronies.
2. IX We recommend both patients and caregivers be educated on proper ventilator use and maintenance.

Respiratory polygraphy or PSG, preferably coupled with transcutaneous capnography, are considered the gold-standard for documenting and correcting undesired respiratory events occurring under HMV.⁷⁹

Unintentional leaks are a major contributor to PVA and side-effects.⁷⁴ Mask related problems (leaks, mucosal irritation, sleep disruption, pressure sores and pain) are common and should be promptly addressed by mask fitting and/or rotation, use of adhesive dressings and medical treatment.^{62,69,74,80} Inappropriate ventilator parameters contribute to leaks, asynchronies, and patient discomfort.^{81,82} PVA represent a mismatch between patient and ventilator, influenced by patient, interface and ventilator factors, potentially leading to ineffective ventilation.^{82–84}

The increasing demand for reliable, accessible and affordable tools to monitor HMV has led to the swift development of technological devices in the most recent decades, mainly in the realm of telemonitoring.^{31,69,71,85,86} Telemonitoring is generally well accepted with cost-effectiveness and improved communication being some of its main advantages.^{87–90} Despite its feasibility and positive effects, evidence on its benefits comparing to in-person follow-up is scarce and its full impact is still uncertain.^{73,74,80,86,89,91–94}

Outcomes

HMV adherence is essential to improve clinical outcomes.⁷³ Data available is scarce but several review articles propose the use of the total number of hours spent on the ventilator, associated with correction of nocturnal hypoventilation as one of the goals in HMV.^{31,73,74,95,96} Based on expert opinion and review of RCTs, several factors could interfere and compromise adherence, namely advanced age, occurrence of comorbidities and cognitive impairment, lack of motivation, treatment delay, inappropriate ventilation pressures and the patients' clinical condition.^{97–100} Intermittent or decreased HMV use may also reflect poor tolerance, discomfort or inappropriate settings and adverse effects.^{71,73}

HRQoL and HMV tolerance may be assessed longitudinally to improve patient-centred outcomes.⁶⁷ Specific patient-reported outcome measures are available for those on HMV such as the Severe Respiratory Insufficiency and the S3-NIV questionnaire.^{101,102} Both have been validated in the Portuguese population.^{103,104}

Therapy withdrawal

There is lack of evidence on the indications for HMV withdrawal. Patient selection seems to be a crucial factor in reducing HMV failure.⁹⁸ Patient's goals, diagnosis, clinical characteristics, and risk of failure should be considered before HMV initiation.⁹⁷ Evidence, mainly based on NMD, suggests that HMV withdrawal should be considered in case of treatment failure or EoL, if the patient so wishes.

Recommendations are presented in Table 3.

Disease specificities

Neuromuscular diseases

Assessment. CRF is a significant concern and a common cause of death in NMD. Non-invasive ventilation (NIV) is recommended for initiating ventilatory support in these patients.^{96,105} Indeed, HMV has increased the survival of patients with NMD, reduced hospital admissions and improved HRQoL and long-term

Table 3. Recommendations for follow-up and monitoring of home mechanical ventilation (HMV).

3.I	We suggest all patients receive follow up within 1 month after starting HMV, and subsequently at 3-to-6-month intervals, according to patients' characteristics and needs.
3.II	Clinical assessment of patients under HMV should focus on addressing symptoms (including dyspnoea, nocturnal hypoventilation symptoms, sleep quality and HMV side effects), assessment of equipment data and its condition, along with patient and caregivers' education.
3.III	We suggest regular monitoring and correction of HMV side-effects (interface-related, ventilator-related and patient-ventilator asynchronies).
3.IV	Healthcare professionals may adopt the use of HMV equipment with incorporated telemonitoring technology, if available and deemed acceptable by both patients and healthcare teams.
3.V	Evidence of factors associated with low adherence, namely patient discomfort or clinical worsening, occurrence of adverse effects, presence of comorbidities and lack of motivation, should be sought for and addressed, to optimize HMV treatment and compliance.
3.VI	HMV withdrawal should be considered on a case-by-case analysis and may be justified by treatment failure, low-adherence, rejection or intolerance despite HMV optimization, significant weight loss leading to resolution of sleep disordered breathing and hypoventilation in patients with OHS and to fulfil end-of-life care decisions.

hypoventilation symptoms.^{106,107} Timely HMV initiation is fundamental when managing patients with NMD.^{40,108,109} Symptoms of respiratory muscle weakness or hypoventilation, if associated with ventilation compromise, are often criteria for HMV namely resting dyspnoea, fatigue, morning headaches, frequent nocturnal awakenings, hypersomnolence, difficulty with concentration and/or irritability, weight loss, but are frequently non-specific findings.¹¹⁰ Dyspnoea and orthopnoea often appear late in the disease course justifying routinely assessing ventilatory and muscular compromise with tests such as slow vital capacity, sitting forced vital capacity (FVC) and supine FVC, muscle pressures – maximum inspiratory pressure (MIP), sniff nasal inspiratory pressure (SNIP) and peak cough flow (PCF), ABG, capnography and eventually sleep studies.^{40,96,111,112} Sleep studies may be required in the presence of symptoms without lung and muscular function compromise.¹¹³

Bulbar disfunction and cough inefficiency. Severe bulbar and glottic dysfunction most commonly occur in patients with ALS, spinal muscle atrophy type 1 and pseudobulbar palsy and carry a negative impact on patient survival and ventilation efficacy.^{114,115} Nutritional optimisation by adjusting food consistencies or gastrostomy placement, Percutaneous endoscopic gastrostomy (PEG) or radiologically inserted gastrostomy (RIG), is frequently needed. In ALS, a greater survival was found in patients where gastrostomy was inserted before significant weight loss.¹¹⁶ Conscious sedation during PEG insertion can be used safely in ALS patients with mild to moderate pulmonary dysfunction.¹¹⁷ In ventilator dependent patients, use of NIV with a nasal mask during PEG or RIG placement was found to be successful and safe.^{118,119}

Difficulty in coughing can signal upcoming ventilation issues and raise the risk of respiratory complications, highlighting the need to implement secretion mobilisation techniques.¹²⁰ It is important to consider manual or mechanical assisted cough, in the presence of compromised cough when PCF < 270 L/min.^{40,121,122}

Follow-up. Due to the progressive and degenerative nature of these diseases, the proposed periodicity of follow-up may vary according to the progression of the disease, from 3 to 6 months.^{40,76} With greater dependence on ventilatory support, it will be necessary to increase the number of hours besides the required adjustments of modes and/or parameters. In the absence of bulbar dysfunction, or when there is a minimal compromise, NIV support, including MPV, effectively and comfortably allow for increased survival and HRQoL.⁴¹ However, in the presence of severe bulbar dysfunction, particularly in ALS, NIV may not be effective despite adequate adjustments, and ventilatory support by tracheostomy should be discussed with the patient and family early in the disease course.^{40,108,113,123}

Recommendations are presented in Table 4.

Table 4. Recommendations for home mechanical ventilation in neuromuscular diseases.

4.I	For patients at risk of respiratory failure, we recommend regular follow-up with clinical and functional evaluation such as slow vital capacity, sitting and supine forced vital capacity (FVC), maximal inspiratory pressure (MIP)/sniff nasal inspiratory pressure (SNIP), maximal expiratory pressure, unassisted and assisted peak cough flow (PCF), diurnal arterial blood gas analysis.
4.II	HMV is suggested in the presence of hypoventilation symptoms and one of the following: – FVC < 50% (in amyotrophic lateral sclerosis FVC < 70%), – MIP < 60 cm H₂O, – SNIP < 40 cm H₂O; – nocturnal hypoventilation, – diurnal hypercapnia.
4.III	Nutritional status and dysphagia should be routinely assessed. Gastrostomy tube placement should be considered in case of significant weight loss or risk of aspiration.
4.IV	Respiratory physiotherapy aimed at enhancing mucociliary clearance, lung volume recruitment and cough efficacy should be available to treat patients with ineffective cough and secretion management.
4.V	Deep lung insufflations to the maximal insufflation capacity combined with manual assisted cough should be applied to patients with unassisted PCF < 270L/min.
4.VI	Mechanical in-exsufflation (MI-E) devices should be considered for patients with assisted PCF < 270L/min and for all patients with invasive ventilatory support.
4.VII	Hypoventilation symptoms, gas exchange and ventilator data analysis are criteria for adjusting ventilatory settings over time and frequency of follow-up can be adjusted depending on the underlying disease and rate of progression.
4.VIII	In the presence of ventilatory dependence (above 12 hours per day) or breathlessness without ventilatory support, daytime ventilation should be proposed, including MPV.
4.IX	Invasive ventilation should be considered in the presence of severe bulbar muscle dysfunction, as non-invasive ventilation and/or MI-E may not be effective despite appropriate adjustments. This should be considered on a case-by-case analysis and decided with the patient.

Obesity-hypoventilation syndrome

Obesity-hypoventilation syndrome is a progressive condition with a high mortality, despite treatment with positive airway pressure (PAP) therapy, with an estimated 3-year mortality of 12–32%.¹²⁴ Patients with OHS often suffer from comorbidities other than obstructive sleep apnoea (OSA), leading to higher mortality and healthcare utilisation and imposing a significant burden on HRQoL.¹²⁵ The impact of obesity in hypoventilation must be perceived as a continuum. Daytime hypercapnia is preceded by hypoventilation during sleep, so daytime hypercapnia already represents an OHS advanced stage.¹²⁶ A staging system was proposed, defining each stage physiopathology characteristics (supplementary material).¹²⁶ Ideally, a staging system would be able to identify which obese patients will develop OHS allowing for prognostication and preventive interventions. Although this proposal needs further validation, there is data supporting that mortality in OHS is higher than in eucapnic obese patients.¹²⁶

Assessment. Evaluation of patients with suspected OHS includes body mass index (BMI) measurement, full blood count, thyroid function, glycosylated haemoglobin, ABG, transcutaneous capnography, echocardiography, PFTs and sleep study.^{126,127}

Patients with known or suspected OSA, who have a serum bicarbonate >27meq/L are likely to have OHS, which should be confirmed with measurement of daytime ABG.¹²⁸ Approximately 90% of patients with OHS have OSA, with nearly 70% having severe OSA. The remaining patients have non-obstructive sleep hypoventilation with no or mild OSA.¹²⁹

We recommend treating OHS with a multimodal approach incorporating PAP therapy, lifestyle changes, physical activity and weight loss to reduce metabolic and cardiovascular risk.¹²⁹ Bariatric surgery is an effective option in severe cases, provided OSA is controlled with PAP.¹²⁵ Glucagon-like peptide 1 receptor agonists show promising results in the reduction of events of patients with OSA and endocrinology referral might be considered.¹³⁰

Positive airway pressure. We recommend patients with OHS be treated with PAP. Stable patients with OHS and concomitant severe OSA should be initially treated with continuous positive airway pressure (CPAP).

Both CPAP and bilevel HMV improve clinical symptoms, polysomnographic parameters, daytime paCO_2 and hospital resource utilisation in patients with OHS and severe OSA.¹³¹ Since CPAP is simpler and more cost-effective than bilevel therapy, CPAP should be the preferred treatment for patients with OHS with severe OSA.¹³² A case-by-case assessment is recommended as the cost-effectiveness advantage may be counterbalanced if patients treated with CPAP have high rates of hospitalisation or hospital resource utilisation.¹³¹

Ideally, initiation of PAP treatment in OHS is set during a titration study performed in a sleep lab or hospital setting.¹²⁹ The OPIP trial showed a similar clinical effectiveness between outpatient using an autotitrating device and inpatient setup strategy.¹³² Due to resource-limitation leading to possible delays in treatment, alternatives to classic PSG-driven titration might be a valuable alternative. In patients with OHS and OSA, several RCTs comparing CPAP to bilevel HMV have failed to find significant differences between these therapies in terms of resolving CRF, improving HRQoL, therapy adherence, incidence of new cardiovascular events or mortality.^{133–136} A proportion of stable OHS patients treated initially with bilevel HMV can be stepped down to CPAP therapy once daytime paCO_2 has normalised.¹³⁷ In OHS patients with left ventricular hypertrophy, pulmonary hypertension and more pure forms of hypoventilation with fewer obstructive events during sleep (i.e., mild to no OSA), bilevel HMV seems to be the treatment of choice.^{129,138}

In patients who remain symptomatic or hypercapnic despite good adherence to CPAP therapy or have significant exacerbations, bilevel HMV should be considered.¹³⁹ However, normalisation of paCO_2 is expected to take longer with CPAP than HMV.^{133,135,136,139}

Using an auto-adjusting PAP device along with sleep study and capnography analysis may be an effective method for short-term titrating treatment in patients with OHS.¹³⁷ Additionally, tools like pulse oximetry and nocturnal transcutaneous capnography are valuable for monitoring and adjusting PAP therapy. After optimisation of positive airway pressure, supplemental oxygen may be added if significant desaturation remains. Oxygen supplementation should be titrated to achieve SpO_2 90–94%, with monitoring for hypercapnia.¹²⁸

Table 5. Recommendations for home mechanical ventilation in obesity-hypoventilation syndrome.

5.I	We recommend a multidisciplinary treatment approach encompassing positive airway pressure (PAP) therapy, lifestyle changes and weight loss.
5.II	We recommend transitioning to non-invasive ventilation (NIV) in patients exhibiting inadequate clinical or blood gas control or experiencing exacerbations while under CPAP.
5.III	If in-laboratory titration is unavailable, outpatient NIV setup may be an alternative in stable patients when performed by experienced healthcare professionals.
5.IV	We suggest close monitoring of PAP therapy in patients with higher body mass index, weight gain, lower FEV ₁ /FVC and need for nocturnal supplemental oxygen, who are particularly prone to treatment failure.

Follow-up. Close monitoring is particularly crucial in patients without severe OSA or those with significant hypercapnia at baseline. Several factors can predict CPAP failure, including severe obesity, weight gain, lower FEV₁/FVC values, and the need for nocturnal supplementary oxygen.¹⁴⁰ Additionally, factors such as diabetes, baseline daytime oxygen saturation below 83%, EPAP less than 7 cmH₂O after titration and adherence to HMV for less than 4 hours can independently predict mortality.¹²⁴

If patients achieve significant weight loss of >10% and symptom improvement, diagnostic workup should be repeated, and treatment change or withdrawal should be considered accordingly.¹⁴¹

Recommendations are presented in Table 5.

Restrictive chest wall diseases

RCTs have not been able to show a survival benefit in this group, however studies have shown HMV can improve HRQoL, decrease hospitalisations, prevent respiratory failure, improve performance and pulmonary haemodynamics.^{12,13,106,107,142–144}

Assessment. We recommend a detailed medical history and physical examination focusing on signs and symptoms of hypoventilation, PFTs (including pletysmography with spirometry, MIP and maximum expiratory pressure, SNIP), diurnal ABG and nocturnal oximetry or transcutaneous capnography.^{12,109,142–146} A sleep study should be performed if concomitant sleep apnoea is suspected.^{12,109,146}

Treatment criteria. Current indications are based on non RCT studies, expert panels and international guidelines.^{12,109,142–146}

Common criteria for initiation are the presence of hypoventilation symptoms in addition to chronic diurnal hypercapnia, nocturnal hypoventilation, and/or FVC < 50%, since international guidelines also highlight the role of hypoventilation symptoms in HMV initiation.^{106,109,142,143,145,146} In the REINVENT survey, the main criteria for initiating long-term HMV in RCWD was diurnal hypercapnia.¹⁴⁶

There is currently no robust evidence to support the use of HMV in patients with nocturnal hypoventilation who do not have diurnal hypercapnia.¹⁰⁶ However, early initiation may be beneficial for patients with FVC < 50% of predicted, who are most likely to develop daytime hypercapnia in the following 2 years.

Treatment specificities. In patients with RCWD, both pressure and volumetric modes have been used.^{60,142,144,145,147} A randomised crossover study comparing these modes found no significant differences in sleep quality, daytime ABG levels, lung mechanics, or daytime functioning.¹⁴⁸ One study compared pressure and volumetric modes, with the former being slightly better tolerated. Low EPAP levels (2–4 cmH₂O) are commonly used except when concomitant OSA is present, in which higher EPAP values should be used.^{142,149} Inspiratory positive airway pressure (IPAP) values typically range around 20 cmH₂O, with backup respiratory rates around 15–16 breaths per minute.^{142,148,149}

Follow up. We suggest revaluation 1 month after HMV initiation, with ABG measurement, and thereafter at 3 to 6-months intervals.¹⁴⁴ Additional adjustments may be needed during follow-up visits related to the patient's symptoms, gas exchange, and tolerance. All patients should be followed-up long-term since the onset of ventilatory failure is often insidious.¹⁴²

Recommendations are presented in Table 6.

Table 6. Recommendations for home mechanical ventilation in restrictive chest wall diseases.

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| 6.I | Both pressure and volume ventilation modes are adequate, but we suggest pressure modes as the first choice (particularly bilevel spontaneous-timed) as they are well tolerated, more widely used and less costly. |
| 6.II | We suggest HMV initiation in the presence of hypoventilation symptoms and any of the following criteria: chronic hypercapnia (PaCO_2 or $\text{tcCO}_2 > 45$ mmHg), nocturnal hypoventilation, oxygen saturation $< 88\%$ for more than five consecutive minutes or oxygen saturation $< 90\%$ during at least 30% of the night, previous admission due to acute hypercapnic respiratory failure requiring NIV, $\text{FVC} < 50\%$ MIP < 60 cmH ₂ O or SNIP < 40 cmH ₂ O. |
| 6.III | When a pressure mode is chosen, we suggest using a low EPAP, unless the patient has concomitant OSA, in which case EPAP levels may be higher (ideally titrated during a sleep study). |
| 6.IV | IPAP levels should provide sufficient pressure support to correct hypoventilation, achieve normocapnia, and eliminate symptoms. |
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COPD

Assessment. COPD and OSA are common interrelated conditions with OSA being particularly prevalent in COPD patients on HMV.^{15,150–152} Although evidence for the performance of the STOP-BANG questionnaire in COPD is absent, it has been found to be the most sensitive screening tool for OSA.¹⁵³ Limited access to diagnostic procedures in COPD patients is currently a major obstacle as PSG is not widely and timely available. Simplified sleep diagnostic methods may increase availability and feasibility.^{154,155} Nocturnal capnography enables the assessment of nocturnal hypoventilation in patients with COPD, and it is suggested, whenever possible, in patients with borderline daytime hypercapnia.¹²

Treatment evidence. In 2014 a RCT by Kohnlein *et al* showed that in severe stable COPD with daytime $\text{paCO}_2 \geq 52$ mmHg the addition of HMV targeted to attain normocapnia (normalising or reducing paCO_2 by at least 20% from baseline), improved survival (for the first time) and HRQoL.⁶³ Also in 2014, the RESCUE trial evaluated COPD patients admitted to the hospital with acute respiratory failure and prolonged hypercapnia defined as $\text{paCO}_2 > 45$ mmHg more than 48 hours after termination of ventilatory support. It concluded that initiation of HMV in the hospital directly after acute hypercapnic respiratory failure, even though it significantly reduced hypercapnia, was not superior regarding patient-important outcomes.¹⁵⁶ In 2017, a RCT by Murphy *et al* investigated the effect of adding HMV to oxygen therapy, among patients with persistent hypercapnia ($\text{paCO}_2 > 53$ mmHg) 2 to 4 weeks after an acute exacerbation of COPD requiring ventilation. The study showed HMV prolonged the time to readmission or death within 12 months.¹⁵⁷ Considering the lack of robust evidence on a single cut-off value of paCO_2 to initiate HMV, we propose using $\text{paCO}_2 \geq 52$ mmHg, which will probably include most patients who may benefit from HMV.^{63,157}

Studies have suggested that in patients with recurrent admissions requiring NIV, HMV can reduce mortality, hospital and intensive care unit admissions, ventilatory needs and costs while improving HRQoL.^{150,158–160} Real-life studies have shown that HMV initiation after acute exacerbation or inability to wean from NIV is common practice.^{15,150,157}

Regarding initiation setting, Duiverman *et al* showed that HMV initiation in stable COPD patients at home using telemedicine is safe, as effective as in-hospital initiation considering paCO_2 reduction and HRQoL improvement with cost reduction.⁵² However, the setup and telemonitoring were performed by experienced clinicians, with close monitoring that mirrored the hospital setting and some groups of patients were excluded such as those post-exacerbation (< 4 weeks), unstable severe cardiac comorbidities or overlap syndrome.⁵² An uncontrolled Portuguese study indicates the outpatient setting was a safe and effective alternative for initiation of HMV in patients with COPD. Patients perceived the experience as positive and had a positive impact on short-term HRQoL.⁶¹

Treatment specificities. A significant reduction or normalisation of paCO_2 is considered fundamental to obtain meaningful outcomes.¹¹ Struik *et al* showed higher improvement of paCO_2 in patients who used HMV for at least 5 hours per night.¹⁶¹

Regarding ventilatory modes, there is no strong evidence that new ventilatory modes are superior to conventional pressure support.¹⁶² Real-life studies demonstrate their usage in a minority of patients.^{4,15} Physiological and clinical benefits of HMV have become more evident since the introduction of 'high-intensity' ventilation, a strategy that is based on titrating pressure support aiming to attain normocapnia, usually requiring high IPAP levels (> 20 cmH₂O) and a high back-up respiratory rate, although the

Table 7. Recommendations for home mechanical ventilation in chronic obstructive pulmonary disease.

7.I	Stable COPD patients with hypercapnia should undergo nocturnal sleep tests to screen for sleep disorder breathing, particularly if symptomatic, obese, with concomitant cardiovascular disease and/or STOP-BANG score ≥ 5 .
7.II	We recommend HMV in patients with persistent daytime $\text{paCO}_2 \geq 52$ mmHg in stable disease or at least 2–4 weeks after an acute exacerbation requiring ventilation.
7.III	HMV could be considered after an acute hypercapnic respiratory failure exacerbation in patients with severe deterioration after a NIV weaning attempt and in patients with recurrent admissions requiring NIV and sustained hypercapnia.
7.IV	We recommend fixed pressure support as the first-choice ventilator mode in patients initiating HMV.
7.V	We recommend nocturnal HMV use for at least 5 hours and titration aiming to normalize or reduce baseline paCO_2 by at least 20%.

additional effect of the latter is debated.^{52,63,163–165} Short rise times (≤ 150 ms) favour a shorter inspiratory time and a favourable I:E ratio (e.g., $\approx 1:3$), improving patient comfort and decreasing work of breathing.¹³

The use of EPAP prevents upper airway collapse during sleep, which is particularly useful in comorbid OSA, and this should be considered when titrating EPAP. Also, in patients with very severe airway obstruction, marked hyperinflation or hypercapnia, higher values of intrinsic positive end expiratory pressure must be anticipated and EPAP values must be titrated accordingly.¹³

Although NIV should not be used routinely during pulmonary rehabilitation, some studies suggest NIV could be considered as an adjuvant therapy in patients on nocturnal HMV, leading to decreased breathlessness and/or higher exercise tolerance.^{166,167}

Recommendations are presented in Table 7.

Other diseases (cystic fibrosis, bronchiectasis, ILD)

Cystic fibrosis and bronchiectasis. In bronchiectasis, evidence is scarce and mainly obtained in patients with cystic fibrosis. Benefits of HMV include improved gas exchange, both awake and asleep, enhanced exercise tolerance, improved muscle strength and better oxygenation during chest physiotherapy.^{168,169} HMV has been shown to stabilise lung function decline in patients with severe lung disease.^{169–171}

For patients on the lung transplant list who suddenly deteriorate with unacceptable hypercapnia and hypoxia, HMV can be utilised as a bridge to transplantation. This is a cost-effective and possibly life-saving intervention and does not increase perioperative complications or survival following lung transplantation.^{172–176}

Currently, there is insufficient data to define clear criteria for HMV initiation in bronchiectasis or cystic fibrosis, as only a limited number of RCT's have been conducted.¹⁷⁷ Use of HMV could be considered in patients with stable persistent diurnal hypercapnia, as adjunct to ACT, or as a bridge to transplantation.^{169,172,173,176,178} In patients with bronchiectasis, humidification and nasal masks may be considered (allowing coughing, secretion clearance and comfort).^{19,176}

Interstitial lung diseases. In ILD, there is scarce data about NIV in acute respiratory failure due to acute exacerbations of idiopathic pulmonary fibrosis but there are no RCTs about the use of HMV in stable ILD with CRF.¹² Patients with ILD are more challenging to ventilate effectively due to lung stiffness from fibrotic changes and frequent nocturnal cough. Nevertheless, a trial of HMV can be considered in selected individuals to determine whether any benefits in terms of gas exchange, symptom relief or HRQoL are achievable.¹³ HMV can be used as a bridge for transplantation, an adjunct to ACT and/or to pulmonary rehabilitation. No specific recommendation about NIV use in pulmonary rehabilitation exists, although some studies had positive results.^{179–181} The benefits of HMV on exacerbation frequency, disease course, sleep quality, and HRQoL have been postulated several times but should also be verified in larger RCTs.^{12,13}

Recommendations are presented in Table 8.

Table 8. Recommendations for home mechanical ventilation in bronchiectasis and interstitial lung diseases.

8.I	Bronchiectasis	HMV might be considered in particular situations, but treatment goals need to be very clear and regularly reevaluated. Consider HMV with humidification for patients with chronic hypercapnic respiratory failure, especially when this is associated with symptoms or recurrent hospitalization, as an adjunct to airway clearance techniques, or as bridge to transplantation.
8.II	ILD	HMV might be considered in particular situations, but treatment goals need to be very clear and regularly reevaluated. Consider HMV in patients with chronic hypercapnic respiratory failure who demonstrate clinical improvement after a HMV trial, as adjunct to respiratory rehabilitation or as bridge to transplantation.

Invasive ventilation

Assessment

Candidates for home mechanical invasive ventilation (HMIV) are a small minority and will remain as a case-by-case selection requiring extensive community resources.^{76,182} HMIV via tracheostomy can ensure adequate ventilation, enhance HRQoL, and reduce hospital admissions but carries a significant burden to patients and caregivers. Despite the lack of RCTs, HMIV in ALS improves survival, but the greatest benefit for this outcome is achieved with previous NIV use.^{183,184}

The discussion for HIMV must emphasise the need for individualised care.^{143,185} Factors such as medical stability, home suitability, caregiver support and training, financial resources, equipment, community healthcare support, and EoL decisions play pivotal roles in the selection criteria for suitable candidates.^{186,187} This discussion should be frank, open and detailed and must include caregivers. Decisions should be documented and accessible to the extended clinical team.

As for most countries, resources in Portugal are unevenly distributed, and there is wide geographical variation that can lead to a postcode lottery regarding access to healthcare, which is true for HMIV as well.¹⁸⁸ As a panel, we acknowledge the challenges but feel much could be improved.

Setting

As a first-line strategy, decreasing the number of patients requiring HMIV would lead to improved patients' outcomes and cost-effectiveness. Literature shows that weaning is successful in around 60 to 80% of patients.^{13,143,182,189,190} Reference centres for difficult weaning provide value by achieving either similar clinical outcomes at a lower cost or better clinical outcomes at a similar cost compared to intensive care units. Importantly, they also offload intensive care units, improving availability for acute cases. We suggest weaning centres be created according to population ratios, receiving referrals from intensive care units or high-dependency respiratory units, and involving multidisciplinary teams with pulmonologists, physiotherapists, speech and language therapists and experienced nursing staff. As a second-line strategy, community support teams for patients unable to be weaned could provide home care and caregiver education. Patients should be selected on a case-by-case analysis, ideally after admission to a weaning centre, with separate interviews for families to consider their perspectives on tracheostomy, and informed conversations regarding the decision-making process.^{185,191,192}

Transitioning patients to HMIV requires meticulous planning, training, and coordination among healthcare teams.⁷⁶ The multidisciplinary approach is a cornerstone of patient care and should address aspects such as nutrition, communication, mobility, social participation, and psychosocial support to better fulfil diverse patients' needs, improving their HRQoL.¹³ Proper caregiver training, encompassing ventilator use, emergency care, and home environment preparation, is essential to ensure the safety and success of HMIV, considering that caregivers are often family members, and play a pivotal role.^{186,193,194}

Treatment specificities

Several aspects of tracheostomy, ventilation equipment, cuff pressure management, cannula management, hygiene, humidification, and minimally invasive suctioning techniques must be mastered.¹⁹⁵ Material selection should consider ease of handling and utility. Formal caregiver training programs are often lacking, underscoring the need for training resources, with special attention to burden and psychological support. Temporary care options could be valuable, as a resource to ease the burden on caregivers.¹⁹⁶ These considerations aim to minimise patient discomfort and complications, ensuring the highest standards of care.

Ventilation mode depends on assessment by specialised pulmonologists. Employing volume-controlled /pressure-controlled ventilation is recommended for patients with progressive conditions or those transitioning from non-invasive to HIMV.¹⁹⁵ A second ventilator and an external battery ensure continuous care for patients with ventilation dependence, allowing patients' mobility.¹⁴³ Adequate alarms for invasive ventilation with life-support ventilators can enhance patients' safety.¹²

A well-trained team should deliver a home primary care-based, tertiary care-based, or a primary-tertiary co-management hybrid model. A physiotherapist or nurse should collaborate in pulmonary rehabilitation whenever needed. Regular follow up with support teams and periodic assessments of caregivers' ability to perform ACT, hygiene routines, material conditions and supplies, and patient stability are crucial for ensuring the patient's well-being.¹⁴³ Proper feedback and reinforcement to patients and caregivers, establishing effective communication between the homecare reality and the hospital clinical team, are also essential.¹⁹⁴

Communication

Communication needs of HMIV patients should be addressed, considering the use of speaking valves and the significance of training for laryngeal function and speech when possible.^{13,197,198} Ventilator pressure and selecting the appropriate cannula that allows airflow to the upper airway when the cuff is deflated is also crucial for successful speech valve application.¹⁹⁹ Importantly, alternative augmentative communication systems, from low-tech to high-tech, should be considered and made available.^{200–202}

Recommendations are presented in [Table 9](#).

Palliative and end of life care

One of the main barriers in patients with CRF is establishing the right time to start palliative care (PC).^{203,204} Needs of patients and their caregivers, surrogate markers of disease severity and/or health service utilisation may help identify those needing PC to avoid prognosis' paralysis.^{64,205} Early and phased introduction of PC should be integrated into ongoing routine care, with generalist/primary PC and when needed with specialist/secondary PC.^{206–208} However, it is still underused in Portugal as shown by a multicentre study in patients with COPD and HMV that revealed that very few patients (4%) were evaluated by a PC team.¹⁵ In a conference paper on the Portuguese ALS HMV Registry, PC follow-up increases to about half of the patients enrolled.²⁰⁹

Advance care planning

Advance care planning (ACP) enables individuals with decisional capacity to define goals and preferences for future medical treatment and care, to discuss these with family and healthcare professionals.^{210,211} It should be started early and reviewed as needed, to avoid crisis-oriented decision-making that increase the burden for surrogates.^{206,207,212} Communication skills are essential to achieve effective and sensitive discussions about goals of care.^{64,211,213} ACP conversations should be documented in the medical record to provide context about the patients' decisions and may include advance directives and assignment of a healthcare proxy.^{214,215} It is fundamental to discuss HMIV early in ACP, respecting patient self-determination, considering advantages and disadvantages, and ideally formalising in advance directives, thereby avoiding emergency tracheostomy.^{216–219}

Table 9. Recommendations for home mechanical invasive ventilation.

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| 9.I | We recommend a multidisciplinary team approach, comprising healthcare professionals, with dedicated staff available for ongoing care, to address the diverse needs of patients, and their caregivers and to cover aspects such as communication, nutrition, mobility and psychosocial support and equitable access. |
| 9.II | We recommend expert assessment of feasibility of HMIV, applying criteria for candidate selection including medical stability, motivation, patients' and family expectations, disclosure of implications, home suitability, caregiver support, financial resources, equipment, training, and community healthcare support. |
| 9.III | We recommend maintaining optimal cuff pressure through regular monitoring, balancing the risks of aspiration and pressure ulcers, favouring cuff-less cannula and/or night-time cuff inflation in patients with preserved bulbar function. We recommend regular training for informal and formal caregivers, as well as healthcare professionals. |
| 9.IV | We recommend individualizing cannula selection based on curvature, diameter, and length, with preference for commercially available flexible cannula materials. We recommend verifying optimal fit by using flexible endoscopy, ensuring the needle tip is at a safe distance from the carina. |
| 9.V | We suggest tailored humidification systems, preferring heated humidifiers over heat-humidity exchangers while at rest and passive moisture filters for diurnal mobility. |
| 9.VI | We recommend promoting non-invasive airway clearance techniques, such as MI-E devices for efficient secretion mobilization. In high-volume/thick mucus production, assisted manual techniques (provided by an healthcare worker or a trained caregiver) should be provided. If needed, MI-E should be combined with minimally invasive suctioning (subglottic aspiration only under direct visualization) and measured tracheal suction depth (1 cm of the tracheostomy cannula) without routine saline installation. |
| 9.VII | We suggest using single-limb non-vented circuits with exhalation valves. |
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Ventilation benefits and burden

Despite HMV benefits, treatment burden on both the patient and their caregivers is often underestimated by healthcare professionals.²²⁰ Reassessing positive and negative impacts should therefore be an active part of follow-up evaluation to elicit the time to change the goals of HMV to be predominantly palliative, i.e., the aim of ventilatory support is to relieve dyspnoea rather than the disease 'itself'.^{221–223} Pharmacological and non-pharmacological treatments should optimise symptomatic control.^{205,206,224} Longitudinal assessment of symptoms and patient-reported outcomes is crucial for disease management.^{207,225}

In EoL, ethical dilemmas can arise, and it is sometimes difficult to decide whether assisted ventilation is improving symptoms or prolonging suffering.^{123,226–228}

EoL care involves providing comprehensive support to patients in their final weeks, days or hours²¹³ Patients have the right to be treated in accordance with their goals of care and not to be subjected to dysthanasia.²²⁹ Their preferred place of care and place of death should be discussed with the patient and family.^{193,213,230,231}

Caregiver burden should be regularly assessed.^{196,205} European PC standards and national legislation recognise their special rights suggesting models of supportive care^{64,229,232–234}

Ventilation withdrawal

Ventilation withdrawal is the discontinuation of mechanical ventilation in a dependent patient, and it is a patient right.^{229,235–238} It shouldn't be confused with medically assisted death, euthanasia or assisted suicide.^{239,240} If the patient is no longer capable of making decisions, ventilation withdrawal can be determined by advance care directives or shared-decision making between family members/healthcare proxy and healthcare multidisciplinary team.²³⁶ Consensus is desirable. However, if agreement cannot be reached, the ethics committee can be involved.²²⁹ A withdrawal plan is crucial for this complex process and the contents to be developed in each phase are set out in supplementary material.

Bereavement

PC offers support to family and other close caregivers during the patient's illness and continues to provide bereavement support, where required, after the patient's death.²⁰³ For a minority of bereaved individuals grieving is complicated justifying the application of a differentiated intervention model.^{241,242}

Recommendations are presented in Table 10.

Future research

One striking characteristic of evidence in the HMV is the overall need for robust data. This is emphasised in most systematic reviews and guidelines. Major limitations in HMV research are the small samples, the impossibility of using a placebo treatment or 'sham' ventilation and therefore inability to conceal patient allocation in RCTs, different practices and patient populations across the world making it difficult for strong evidence-based results.

Table 10. Recommendations for the integration of palliative and end-of-life care in home mechanical ventilation.

10.I	We suggest treatment goals prioritize the reduction of symptom burden and improvement of HRQoL through the longitudinal measurement of short and simple patient-reported questionnaires.
10.II	We recommend optimizing physical and psychological symptom control with primary palliative actions or secondary palliative care according to the needs of each patient, their family and social context.
10.III	We recommend adopting a person-centred approach when engaging in Advanced Care Planning (ACP) conversations. Patient-designated surrogates or healthcare proxies should be included across all stages with patient permission. Preferences should be reviewed whenever circumstances change.
10.IV	We suggest regularly discussing treatment burden when palliative HMV is ongoing. To maximize comfort and effectiveness (symptom and suffering reduction) the follow-up must be tailored to the patient and their caregivers' needs and wishes, and adapted to local resources, until the end of life.
10.V	If NIV is no longer tolerated or effective, ACP review is crucial to decide for HMIV or palliation of symptoms.
10.VI	We suggest including an early evaluation of the patient caregivers' needs and wishes in the HMV care plan.
10.VII	We suggest using a ventilation withdrawal protocol/check list adjusted to local resources and organization, in patients dependent of HMV that no longer desire to keep ventilation support. Five key steps should be addressed in the protocol: 1) Decision making, 2) Communication, 3) Withdrawal planning, 4) Withdrawal and 5) Bereavement.
10. VIII	We recommend that healthcare professionals involve caregivers and family members in end-of-life care, support them to prevent Prolonged Grief Disorder and guide them according to best clinical practices

We believe that adequately designed and executed international RCTs with larger samples are still needed, particularly on the following areas:

- Determining which outcomes are more adequate and valued to measure HMV effectiveness;
- Identifying accurate criteria for selecting patients likely to benefit from long-term HMV;
- Comparing different initiation and follow-up settings and protocols in terms of efficacy, safety, costs and patient preference;
- Evaluating if automated ventilatory modes offer advantage over traditional modes;
- Assessing patient-reported outcomes and experience measures, and caregiver burden related to treatment;
- Analysing cost effectiveness and patient/family costs of treatment;
- Determining what will be the impact technological advances, such as telemedicine, on treatment initiation and follow up.

Conclusion

With this document, the HMV assembly of the Portuguese Respiratory Society aims to provides clinical practice recommendations for HMV adapted to the Portuguese healthcare system, highlights current evidence gaps, and provides suggestions for future research.

Acknowledgments

The authors would like to thank Begum Ergan, Claudia Crimi, Jean Paul Janssens, Manuel Lujan, Marieke Duiverman, Maxime Patout, Ove Fondenes, Paola Pierucci and Tiina Anderson for contributing to the second round of the Delphi process and Anita Simonds, Jean Paul Janssens, and Paola Pierucci for proofreading the manuscript.

Disclosure statement

Ana Cysneiros declares to have received non-financial support (support for attending meetings and/or travel) from Linde Saúde. Carla Ribeiro declares to have received expert testimony/lecture fees from Nippon Gases and Vitalaire and non-financial support (support for attending meetings and/or travel) from Linde Saúde, Nippon Gases, Vitalaire and Acail Gás, all outside the submitted proposal. Lucia Mendez Gonzalez declares to have received consulting fees from Boehringer Ingelheim, CSL Behring and Nippon Gases, all outside the submitted proposal. No potential competing interest was reported by: Alexandra Carreiro, Ana Luísa Vieira, Bárbara Seabra, Bebiana Conde, Carla Nogueira, Célia Durães, Cidália Rodrigues, Cláudia Pimenta, Cristina Cristóvão, Cristina Jácome, Daniela Rodrigues, Diva Ferreira, Filipe Gonçalves, Helena Chaves Ramos, Joana Lages, João Cravo, João Paulo Silva, Karl Cunha, Mafalda Van Zeller, Márcia Araujo, Margarida Aguiar, Margarida Barata, Margarida Raposo, Margarida Redondo, Maria Jacob, Maria João Araújo, Marta Drummond, Miguel Gonçalves, Miguel Guia, Monica Grafino, Nuno Faria, Paula Pamplona, Paula Pinto, Pedro Viegas, Rita Gomes, Sara Conde

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Data availability statement

Data not available due to nature of the research.

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