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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.

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Home mechanical ventilation; recommendations; clinical practice; expert consensus

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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 heterogenicity 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 bimonthly 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 ($CO_2$)-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 ($tCO_2$). 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:<br>– FVC < 50% (in amyotrophic lateral sclerosis FVC < 70%),<br>– MIP < 60 cm $H_2O$,<br>– SNIP < 40 cm $H_2O$;<br>– nocturnal hypoventilation,<br>– 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, glycated 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 dessaturation 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 $FEV_1$/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 $cmH_2O$ 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 $cmH_2O$) 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 $cmH_2O$, 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.

| | |
|---|---|
| 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 $tcCO_2$ >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, FVC < 50% MIP < 60 $cmH_2O$ or SNIP < 40 $cmH_2O$. |
| 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. |

## *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 $paCO_2$ ≥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 $paCO_2$ > 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 ($paCO_2$ > 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 $paCO_2$ ≥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 specificitie.*** 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 $cmH_2O$) 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 $paCO_2$ ≥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., ≈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 transplantion, 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.

| | |
|---|---|
| 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. |

### *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 hours[213] 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.

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## Data availability statement

Data not available due to nature of the research.

## References

1. Patout M, Lhuillier E, Kaltsakas G, et al. Long-term survival following initiation of home non-invasive ventilation: a European study. *Thorax*. 2020;75(11):965–973. doi: 10.1136/thoraxjnl-2019-214204.
2. Schwarz EI, Mackie M, Weston N, et al. Time-to-death in chronic respiratory failure on home mechanical ventilation: a cohort study. *Respir Med*. 2020;162:105877. doi: 10.1016/j.rmed.2020.105877.
3. Toussaint M, Wijkstra PJ, McKim D, et al. Building a home ventilation programme: population, equipment, delivery and cost. *Thorax*. 2022;77(11):1140–1148. doi: 10.1136/thoraxjnl-2021-218410.
4. Cantero C, Adler D, Pasquina P, et al. Long-term noninvasive ventilation in the Geneva Lake area: indications, prevalence, and modalities. *Chest*. 2020;158(1):279–291. doi: 10.1016/j.chest.2020.02.064.
5. Ribeiro Baptista B, Baptiste A, Granger B, et al. Growth of home respiratory equipment from 2006 to 2019 and cost control by health policies. *Respir Med Res*. 2022;82:100930. doi: 10.1016/j.resmer.2022.100930.
6. Lloyd-Owen SJ, Donaldson GC, Ambrosino N, et al. Patterns of home mechanical ventilation use in Europe: results from the Eurovent survey. *Eur Respir J*. 2005;25(6):1025–1031. doi: 10.1183/09031936.05.00066704.
7. Fundação Portuguesa do Pulmão. Observatório Nacional Doenças Respiratórias. 2023. https://www.fundacaoportuguesadopulmao.org/destaques/conteudos/ondr-2023/.
8. Melloni B, Mounier L, Laaban JP, Chambellan A, Foret D, Muir JF. Home-based care evolution in chronic respiratory failure between 2001 and 2015 (Antadir Federation Observatory). *Respiration*. 2018;96(5):446–454. doi: 10.1159/000490549.
9. Winck JC. Normas e Recomendações para a Ventiloterapia Domiciliária - Relatório do sub-grupo de Ventiloterapia Domiciliária da Comissão de Trabalho de Reabilitação Respiratória. *Rev Port Pneumol*. 1998;4(5):447–478. doi: 10.1016/S0873-2159(15)31068-0.
10. Direcção Geral de Saúde. Cuidados Respiratórios Domiciliários: Prescrição de Ventiloterapia e outros equipamentos. 2011. [updated 2015]. https://www.dgs.pt/directrizes-da-dgs/normas-e-circulares-normativas/norma-n-0222011-de-28092011-jpg.asp.
11. Ergan B, Oczkowski S, Rochwerg B, et al. European Respiratory Society guidelines on long-term home non-invasive ventilation for management of COPD. *Eur Respir J*. 2019;54(3):1901003. doi: 10.1183/13993003.01003-2019.
12. Windisch W, Geiseler J, Simon K, Walterspacher S, Dreher M, On Behalf of the Guideline C. German national guideline for treating chronic respiratory failure with invasive and non-invasive ventilation - revised edition 2017: part 2. *Respiration*. 2018;96(2):171–203.
13. Janssens JP, Michel F, Schwarz EI, et al. Long-term mechanical ventilation: recommendations of the Swiss Society of Pulmonology. *Respiration*. 2020:1–36. doi: 10.1159/000510086.
14. Serviços Partilhados Ministério da Saúde. Caderno de Encargos - Procedimento de contratação tendente à celebração de acordo quadro para a área da saúde com vista à prestação de Serviços de Cuidados Técnicos Respiratórios Domiciliários CP 131/2022. 2024. https://www.spms.min-saude.pt/wp-content/uploads/2024/10/CE_CRD_2republicacao.pdf.
15. Ribeiro C, Vieira AL, Pamplona P, et al. Current practices in home mechanical ventilation for chronic obstructive pulmonary disease: a real-life cross-sectional multicentric study. *Int J Chron Obstruct Pulmon Dis*. 2021;16:2217–2226. doi: 10.2147/COPD.S314826.
16. Mineiro MA, Guimarães MJ, Winck JC. Organization of home mechanical ventilation in Portugal: characterization of current centers and a pathway to uniformization. *Pulmonology*. 2020;26(2):84–89. doi: 10.1016/j.pulmoe.2019.05.006.

17. Gregoretti C, Navalesi P, Ghannadian S, Carlucci A, Pelosi P. Choosing a ventilator for home mechanical ventilation. *Breathe*. 2013;9(5):394–409. doi: 10.1183/20734735.042312.
18. Falaize L, Leroux K, Prigent H, et al. Battery life of portable home ventilators: effects of ventilator settings. *Respir Care*. 2014;59(7):1048–1052. doi: 10.4187/respcare.02711.
19. Simonds AK, ed. *ERS Practical Handbook of Noninvasive Ventilation*. Lausanne, Switzerland: European Respiratory Society (ERS); 2015:320. doi: 10.1183/9781849840767.eph01.
20. Toussaint M, Reychler G. Recommendations for hygiene of masks and circuits in mechanically home ventilated patients. *Braz J Infect Dis*. 2010;14(4):380–384. doi: 10.1016/S1413-8670(10)70080-5.
21. Rodríguez González-Moro JM, Andrade Vivero G, de Miguel Díez J, et al. Bacterial colonization and home mechanical ventilation: prevalence and risk factors. *Arch Bronconeumol*. 2004;40(9):392–396. doi: 10.1016/S0300-2896(04)75557-X.
22. Singh A, Sterk PJ. Noninvasive ventilation and the potential risk of transmission of infection. *Eur Respir J*. 2008;32(3):816. doi: 10.1183/09031936.00052208.
23. Crescimanno G, Marrone O, Vianello A. Efficacy and comfort of volume-guaranteed pressure support in patients with chronic ventilatory failure of neuromuscular origin. *Respirology*. 2011;16(4):672–679. doi: 10.1111/j.1440-1843.2011.01962.x.
24. Nilius G, Katamadze N, Domanski U, Schroeder M, Franke KJ. Non-invasive ventilation with intelligent volume-assured pressure support versus pressure-controlled ventilation: effects on the respiratory event rate and sleep quality in COPD with chronic hypercapnia. *Int J Chron Obstruct Pulmon Dis*. 2017;12:1039–1045. doi: 10.2147/COPD.S126970.
25. Struik FM, Duiverman ML, Meijer PM, Nieuwenhuis JA, Kerstjens HA, Wijkstra PJ. Volume-targeted versus pressure-targeted noninvasive ventilation in patients with chest-wall deformity: a pilot study. *Respir Care*. 2011;56(10):1522–1525. doi: 10.4187/respcare.01043.
26. Patout M, Gagnadoux F, Rabec C, et al. Avaps-ae versus ST mode: a randomized controlled trial in patients with obesity hypoventilation syndrome. *Respirology*. 2020;25(10):1073–1081. doi: 10.1111/resp.13784.
27. Windisch W, Storre JH, Sorichter S, Virchow JC. Comparison of volume- and pressure-limited NPPV at night: a prospective randomized cross-over trial. *Respir Med*. 2005;99(1):52–59. doi: 10.1016/j.rmed.2004.05.009.
28. Gonzalez-Bermejo J, Nava S, Rabec C, Vega ML. Precision medicine is coming to town: personalising home ventilatory equipment in COPD patients with chronic hypercapnic respiratory failure. *Eur Respir Rev*. 2019;28(154):190043. doi: 10.1183/16000617.0043-2019.
29. Farré R, Navajas D, Montserrat JM. Technology for noninvasive mechanical ventilation: looking into the Black box. *ERJ Open Res*. 2016;2(1):00004–2016. doi: 10.1183/23120541.00004-2016.
30. Carlucci A, Patout M, Winck JC. Does one size fit all? An update on chronic ventilatory support in different respiratory illnesses. *Breathe (Sheff)*. 2023;19(2):230046. doi: 10.1183/20734735.0046-2023.
31. Duiverman ML. "Tricks and tips for home mechanical ventilation" home mechanical ventilation: set-up and monitoring protocols. *Pulmonology*. 2021;27(2):144–150. doi: 10.1016/j.pulmoe.2020.08.002.
32. Khirani S, Ramirez A, Delord V, et al. Evaluation of ventilators for mouthpiece ventilation in neuromuscular disease. *Respir Care*. 2014;59(9):1329–1337. doi: 10.4187/respcare.03031.
33. Ogna A, Prigent H, Falaize L, et al. Accuracy of tidal volume delivered by home mechanical ventilation during mouthpiece ventilation: a bench evaluation. *Chron Respir Dis*. 2016;13(4):353–360. doi: 10.1177/1479972316647177.
34. Masefield S, Vitacca M, Dreher M, et al. Attitudes and preferences of home mechanical ventilation users from four European countries: an ERS/ELF survey. *ERJ Open Res*. 2017;3(2):00015–2017. doi: 10.1183/23120541.00015-2017.
35. McArdle N, Rea C, King S, et al. Treating chronic hypoventilation with automatic adjustable versus fixed EPAP intelligent volume-assured positive airway pressure support (iVAPS): a randomized controlled trial. *Sleep*. 2017;40(10). doi:10.1093/sleep/zsx136.
36. Arellano-Maric MP, Gregoretti C, Duiverman M, Windisch W. Long-term volume-targeted pressure-controlled ventilation: sense or nonsense? *Eur Respir J*. 2017;49(6):1602193. doi: 10.1183/13993003.02193-2016.
37. Garuti G, Nicolini A, Grecchi B, Lusuardi M, Winck JC, Bach JR. Open circuit mouthpiece ventilation: concise clinical review. *Pulmonology*. 2014;20(4):211–218. doi: 10.1016/j.rppneu.2014.03.004.
38. Lofaso F, Prigent H, Tiffreau V, et al. Long-term mechanical ventilation equipment for neuromuscular patients: meeting the expectations of patients and prescribers. *Respir Care*. 2014;59(1):97–106. doi: 10.4187/respcare.02229.
39. Toussaint M, Chatwin M, Verhulst S, Reychler G. Preference of neuromuscular patients regarding equipment for daytime mouthpiece ventilation: a randomized crossover study. *Clin Respir J*. 2020;14(3):214–221. doi: 10.1111/crj.13118.
40. Khan A, Frazer-Green L, Amin R, et al. Respiratory management of patients with neuromuscular weakness: an American College of Chest Physicians clinical practice guideline and expert panel report. *Chest*. 2023;164(2):394–413. doi: 10.1016/j.chest.2023.03.011.
41. Toussaint M, Chatwin M, Gonçalves MR, et al. Mouthpiece ventilation in neuromuscular disorders: narrative review of technical issues important for clinical success. *Respir Med*. 2021;180:106373. doi: 10.1016/j.rmed.2021.106373.
42. Nava S, Navalesi P, Gregoretti C. Interfaces and humidification for noninvasive mechanical ventilation. *Respir Care*. 2009;54(1):71–84.

43. Pierucci P, Portacci A, Carpagnano GE, et al. The right interface for the right patient in noninvasive ventilation: a systematic review. *Expert Rev Respir Med*. 2022;16(8):931–944. doi: 10.1080/17476348.2022.2121706.
44. Szkulmowski Z, Belkhouja K, Le QH, Robert D, Argaud L. Bilevel positive airway pressure ventilation: factors influencing carbon dioxide rebreathing. *Intensive Care Med*. 2010;36(4):688–691. doi: 10.1007/s00134-010-1774-z .
45. Schettino GP, Chatmongkolchart S, Hess DR, Kacmarek RM. Position of exhalation port and mask design affect CO2 rebreathing during noninvasive positive pressure ventilation. *Crit Care Med*. 2003;31(8):2178–2182.
46. Storre JH, Huttmann SE, Ekkernkamp E, et al. Oxygen supplementation in noninvasive home mechanical ventilation: the crucial roles of CO2 exhalation systems and leakages. *Respir Care*. 2014;59(1):113–120. doi: 10.4187/respcare.02596.
47. Lellouche F, Maggiore SM, Lyazidi A, Deye N, Taillé S, Brochard L. Water content of delivered gases during non-invasive ventilation in healthy subjects. *Intensive Care Med*. 2009;35(6):987–995. doi: 10.1007/s00134-009-1455-y.
48. Restrepo RD, Walsh BK. Humidification during invasive and noninvasive mechanical ventilation: 2012. *Respir Care*. 2012;57(5):782–788. doi: 10.4187/respcare.01766.
49. Holland AE, Denehy L, Buchan CA, Wilson JW. Efficacy of a heated passover humidifier during noninvasive ventilation: a bench study. *Respir Care*. 2007;52(1):38–44.
50. Collada-Carrasco J, Lamolda-Puyol C, Luján M, et al. The addition of a humidifier device to a circuit and its impact on home ventilator performance: a bench study. *Pulmonology*. 2020;26(6):363–369. doi: 10.1016/j.pulmoe.2019.11.004.
51. Kampelmacher MJ. Moving from inpatient to outpatient or home initiation of non-invasive home mechanical ventilation. *J Clin Med*. 2023;12(8):2981. doi: 10.3390/jcm12082981.
52. Duiverman ML, Vonk JM, Bladder G, et al. Home initiation of chronic non-invasive ventilation in COPD patients with chronic hypercapnic respiratory failure: a randomised controlled trial. *Thorax*. 2020;75(3):244–252. doi: 10.1136/thoraxjnl-2019-213303.
53. van den Biggelaar RJM, Hazenberg A, Cobben NAM, Gaytant MA, Vermeulen KM, Wijkstra PJ. A randomized trial of initiation of chronic noninvasive mechanical ventilation at home vs in-hospital in patients with neuromuscular disease and thoracic cage disorder: the Dutch HOMERUN trial. *Chest*. 2020;158(6):2493–2501. doi: 10.1016/j.chest.2020.07.007.
54. Lujan M, Moreno A, Veigas C, Monton C, Pomares X, Domingo C. Non-invasive home mechanical ventilation: effectiveness and efficiency of an outpatient initiation protocol compared with the standard in-hospital model. *Respir Med*. 2007;101(6):1177–1182. doi: 10.1016/j.rmed.2006.11.006.
55. Chatwin M, Nickol AH, Morrell MJ, Polkey MI, Simonds AK. Randomised trial of inpatient versus outpatient initiation of home mechanical ventilation in patients with nocturnal hypoventilation. *Respir Med*. 2008;102(11):1528–1535. doi: 10.1016/j.rmed.2008.07.019.
56. Sheers N, Berlowitz DJ, Rautela L, Batchelder I, Hopkinson K, Howard ME. Improved survival with an ambulatory model of non-invasive ventilation implementation in motor neuron disease. *Amyotroph Lateral Scler Frontotemporal Degener*. 2014;15(3–4):180–184. doi: 10.3109/21678421.2014.881376.
57. Bertella E, Banfi P, Paneroni M, et al. Early initiation of night-time NIV in an outpatient setting: a randomized non-inferiority study in ALS patients. *Eur J Phys Rehabil Med*. 2017;53(6):892–899. doi: 10.23736/S1973-9087.17.04511-7.
58. Volpato E, Vitacca M, Ptacinsky L, et al. Home-based adaptation to night-time non-invasive ventilation in patients with amyotrophic lateral sclerosis: a randomized controlled trial. *J Clin Med*. 2022;11(11):3178. doi: 10.3390/jcm11113178.
59. Domenech-Clar R, Nauffal-Manssur D, Compte-Torrero L, Rosales-Almazan MD, Martinez-Perez E, Soriano-Melchor E. Adaptation and follow-up to noninvasive home mechanical ventilation: ambulatory versus hospital. *Respir Med*. 2008;102(11):1521–1527. doi: 10.1016/j.rmed.2008.07.018.
60. Pallero M, Puy C, Güell R, et al. Ambulatory adaptation to noninvasive ventilation in restrictive pulmonary disease: a randomized trial with cost assessment. *Respir Med*. 2014;108(7):1014–1022. doi: 10.1016/j.rmed.2014.04.016.
61. Ribeiro C, Jácome C, Oliveira P, Luján M, Conde S. Impact of outpatient adaptation to home mechanical ventilation on health-related quality of life in patients with chronic obstructive pulmonary disease: the OutVent study. *ERJ Open Res*. 2024;10(5):00125–2024. doi: 10.1183/23120541.00125-2024.
62. Ribeiro C, Jácome C, Oliveira P, Conde S, Windisch W, Nunes R. Patients experience regarding home mechanical ventilation in an outpatient setting. *Chron Respir Dis*. 2022;19:14799731221137082. doi: 10.1177/14799731221137082.
63. Köhnlein T, Windisch W, Köhler D, et al. Non-invasive positive pressure ventilation for the treatment of severe stable chronic obstructive pulmonary disease: a prospective, multicentre, randomised, controlled clinical trial. *Lancet Respir Med*. 2014;2(9):698–705. doi: 10.1016/S2213-2600(14)70153-5.
64. Janssen DJA, Bajwah S, Boon MH, et al. European Respiratory Society clinical practice guideline: palliative care for people with COPD or interstitial lung disease. *Eur Respir J*. 2023;62(2):2202014. doi: 10.1183/13993003.02014-2022.
65. Hannan LM, Rautela L, Berlowitz DJ, et al. Randomised controlled trial of polysomnographic titration of non-invasive ventilation. *Eur Respir J*. 2019;53(5):1802118. doi: 10.1183/13993003.02118-2018.

66. Patout M, Arbane G, Cuvelier A, Muir JF, Hart N, Murphy PB. Polysomnography versus limited respiratory monitoring and nurse-led titration to optimise non-invasive ventilation set-up: a pilot randomised clinical trial. *Thorax*. 2019;74(1):83–86. doi: 10.1136/thoraxjnl-2017-211067.
67. Jolly G, Razakamanantsoa L, Fresnel E, Gharsallaoui Z, Cuvelier A, Patout M. Defining successful non-invasive ventilation initiation: data from a real-life cohort. *Respirology*. 2021;26(11):1067–1075. doi: 10.1111/resp.14118.
68. Elliott MW. Non-invasive ventilation during sleep: time to define new tools in the systematic evaluation of the technique. *Thorax*. 2011;66(1):82–84. doi: 10.1136/thx.2010.142117.
69. Arnal JM, Texereau J, Garnero A. Practical insight to monitor home NIV in COPD patients. *COPD*. 2017;14 (4):401–410. doi: 10.1080/15412555.2017.1298583.
70. Chiner E, Sancho-Chust JN, Landete P, Senent C, Gómez-Merino E. Complementary home mechanical ventilation techniques. SEPAR year 2014. *Arch Bronconeumol*. 2014;50(12):546–553.
71. van der Leest S, Duiverman ML. High-intensity non-invasive ventilation in stable hypercapnic COPD: evidence of efficacy and practical advice. *Respirology*. 2019;24(4):318–328. doi: 10.1111/resp.13450.
72. Janssens JP, Borel JC, Pepin JL, Somno NIVG. Nocturnal monitoring of home non-invasive ventilation: the contribution of simple tools such as pulse oximetry, capnography, built-in ventilator software and autonomic markers of sleep fragmentation. *Thorax*. 2011;66(5):438–445. doi: 10.1136/thx.2010.139782.
73. Borel JC, Palot A, Patout M. Technological advances in home non-invasive ventilation monitoring: reliability of data and effect on patient outcomes. *Respirology*. 2019;24(12):1143–1151. doi: 10.1111/resp.13497.
74. Janssens JP, Cantero C, Pasquina P, Georges M, Rabec C. Monitoring long term noninvasive ventilation: benefits, caveats and perspectives. *Front Med (Lausanne)*. 2022;9:874523. doi: 10.3389/fmed.2022.874523.
75. Crimi C, Noto A, Princi P, et al. Domiciliary non-invasive ventilation in COPD: an international survey of indications and practices. *COPD*. 2016;13(4):483–490. doi: 10.3109/15412555.2015.1108960 .
76. Georges M, Perez T, Rabec C, et al. Proposals from a French expert panel for respiratory care in ALS patients. *Respir Med Res*. 2022;81:100901. doi: 10.1016/j.resmer.2022.100901.
77. Leger P, Laier-Groeneveld G. Infrastructure, funding and follow-up in a programme of noninvasive ventilation. *Euro Respir J*. 2002;20(6):1573–1578. doi: 10.1183/09031936.02.00405802.
78. Berry RB, Budhiraja R, Gottlieb DJ, et al. Rules for scoring respiratory events in sleep: update of the 2007 AASM manual for the scoring of sleep and associated events. Deliberations of the sleep apnea definitions task force of the American Academy of Sleep Medicine. *J Clin Sleep Med*. 2012;8(5):597–619. doi: 10.5664/jcsm.2172.
79. Gonzalez-Bermejo J, Perrin C, Janssens JP, et al. Proposal for a systematic analysis of polygraphy or polysomnography for identifying and scoring abnormal events occurring during non-invasive ventilation. *Thorax*. 2012;67 (6):546–552. doi: 10.1136/thx.2010.142653.
80. Arnal JM, Oranger M, Gonzalez-Bermejo J. Monitoring systems in home ventilation. *J Clin Med*. 2023;12(6):2163. doi: 10.3390/jcm12062163.
81. Hill NS. Noninvasive ventilation in chronic obstructive pulmonary disease. *Clin Chest Med*. 2000;21(4):783–797. doi: 10.1016/S0272-5231(05)70184-2.
82. Bello G, De Pascale G, Antonelli M. Noninvasive ventilation. *Clin Chest Med*. 2016;37(4):711–721. doi: 10.1016/j.ccm.2016.07.011.
83. Longhini F, Bruni A, Garofalo E, et al. Monitoring the patient-ventilator asynchrony during non-invasive ventilation. *Front Med (Lausanne)*. 2022;9:1119924. doi: 10.3389/fmed.2022.1119924.
84. Al Otair HA, BaHammam AS. Ventilator- and interface-related factors influencing patient-ventilator asynchrony during noninvasive ventilation. *Ann Thorac Med*. 2020;15(1):1–8. doi: 10.4103/atm.ATM_24_19.
85. Ackrivo J, Elman L, Hansen-Flaschen J. Telemonitoring for home-assisted ventilation: a narrative review. *Ann Am Thorac Soc*. 2021;18(11):1761–1772. doi: 10.1513/AnnalsATS.202101-033CME.
86. Duiverman ML, Ribeiro C, Tonia T, et al. European Respiratory Society clinical practice guideline on telemedicine in home mechanical ventilation. *Eur Respir J*. 2025;66(5):2500094. doi: 10.1183/13993003.00094-2025.
87. Lopes de Almeida JP, Pinto A, Pinto S, Ohana B, de Carvalho M. Economic cost of home-telemonitoring care for BiPAP-assisted ALS individuals. *Amyotroph Lateral Scler*. 2012;13(6):533–537. doi: 10.3109/17482968.2012.703675.
88. de Almeida JP, Pinto AC, Pereira J, Pinto S, de Carvalho M. Implementation of a wireless device for real-time telemedical assistance of home-ventilated amyotrophic lateral sclerosis patients: a feasibility study. *Telemed J E Health*. 2010;16(8):883–888.
89. Baltaxe E, Embid C, Aumatell E, et al. Integrated care intervention supported by a mobile health tool for patients using noninvasive ventilation at home: randomized controlled trial. *JMIR Mhealth Uhealth*. 2020;8(4):e16395. doi: 10.2196/16395.
90. Ando H, Ashcroft-Kelso H, Halhead R, et al. Experience of telehealth in people with motor neurone disease using noninvasive ventilation. *Disabil Rehabil Assist Technol*. 2021;16(5):490–496. doi: 10.1080/17483107.2019.1659864.
91. Jiang W, Wang L, Song Y. Titration and follow-up for home noninvasive positive pressure ventilation in chronic obstructive pulmonary disease: the potential role of tele-monitoring and the Internet of things. *Clin Respir J*. 2021;15(7):705–715. doi: 10.1111/crj.13352.
92. Ambrosino N, Pierucci P. Using telemedicine to monitor the patient with chronic respiratory failure. *Life (Basel)*. 2021;11(11):1113. doi: 10.3390/life11111113.

93. Ambrosino N, Fracchia C. The role of tele-medicine in patients with respiratory diseases. *Expert Rev Respir Med*. 2017;11(11):893–900. doi: 10.1080/17476348.2017.1383898.
94. Pontier-Marchandise S, Texereau J, Prigent A, et al. Home NIV treatment quality in patients with chronic respiratory failure having participated to the French nationwide telemonitoring experimental program (the TELVENT study). *Respir Med Res*. 2023;84:101028. doi: 10.1016/j.resmer.2023.101028.
95. Rabec C, Rodenstein D, Leger P, et al. Ventilator modes and settings during non-invasive ventilation: effects on respiratory events and implications for their identification. *Thorax*. 2011;66(2):170–178. doi: 10.1136/thx.2010.142661.
96. Hansen-Flaschen J, Ackrivo J. Practical guide to management of long-term noninvasive ventilation for adults with chronic neuromuscular disease. *Respir Care*. 2023;68(8):1123–1157. doi: 10.4187/respcare.10349.
97. Hill NS. Noninvasive ventilation for chronic obstructive pulmonary disease. *Respir Care*. 2004;49(1):72–87; discussion–9.
98. Carron M, Freo U, BaHammam AS, et al. Complications of non-invasive ventilation techniques: a comprehensive qualitative review of randomized trials. *Br J Anaesth*. 2013;110(6):896–914. doi: 10.1093/bja/aet070.
99. Gigante AR, Tinoco EM, Nogueira C, Conde S, Ribeiro C. Factors influencing non-acceptance of home non-invasive ventilation. *Respir Med Res*. 2022;82:100938. doi: 10.1016/j.resmer.2022.100938.
100. Patout M, Razakamanantsoa L, D'Cruz R, et al. Cognitive function in patients with chronic respiratory failure: effect of treatment with home noninvasive ventilation. *Am J Respir Crit Care Med*. 2023;208(8):900–903. doi: 10.1164/rccm.202212-2323LE.
101. Windisch W, Freidel K, Schucher B, et al. The severe respiratory insufficiency (SRI) questionnaire: a specific measure of health-related quality of life in patients receiving home mechanical ventilation. *J Clin Epidemiol*. 2003;56(8):752–759. doi: 10.1016/S0895-4356(03)00088-X.
102. Dupuis-Lozeron E, Gex G, Pasquina P, et al. Development and validation of a simple tool for the assessment of home noninvasive ventilation: the S 3 -NIV questionnaire. *Eur Respir J*. 2018;52(5):1801182. doi: 10.1183/13993003.011822018.
103. Ribeiro C, Ferreira D, Conde S, Oliveira P, Windisch W. Validation of the Portuguese severe respiratory insufficiency questionnaire for home mechanically ventilated patients. *Rev Port Pneumol (2006)*. 2017;23(3):139–145. doi: 10.1016/j.rppnen.2017.01.001.
104. Ribeiro C, Conde S, Oliveira P, et al. Portuguese adaptation of the S3-non-invasive ventilation (S3-NIV) questionnaire for home mechanically ventilated patients. *Pulmonology*. 2020;28(4):262–267. doi: 10.1016/j.pulmoe.2020.11.006.
105. Bourke SC, Tomlinson M, Williams TL, Bullock RE, Shaw PJ, Gibson GJ. Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: a randomised controlled trial. *Lancet Neurol*. 2006;5(2):140–147. doi: 10.1016/S1474-4422(05)70326-4.
106. Annane D, Orlikowski D, Chevret S. Nocturnal mechanical ventilation for chronic hypoventilation in patients with neuromuscular and chest wall disorders. *Cochrane Database Syst Rev*. 2014;2014(12):CD001941. doi: 10.1002/14651858.CD001941.pub3.
107. Hannan LM, Dominelli GS, Chen YW, Darlene Reid W, Road J. Systematic review of non-invasive positive pressure ventilation for chronic respiratory failure. *Respir Med*. 2014;108(2):229–243. doi: 10.1016/j.rmed.2013.11.010.
108. Morelot-Panzini C, Bruneteau G, Gonzalez-Bermejo J. NIV in amyotrophic lateral sclerosis: the 'when' and 'how' of the matter. *Respirology*. 2019;24(6):521–530. doi: 10.1111/resp.13525.
109. Wolfe LF, Benditt JO, Aboussouan L, Hess DR, Coleman JM 3rd. Optimal NIV medicare access promotion: patients with thoracic restrictive disorders: a technical expert panel report from the American College of Chest Physicians, the American Association for Respiratory Care, the American Academy of Sleep Medicine, and the American Thoracic Society. *Chest*. 2021;160(5):e399–e408. doi: 10.1016/j.chest.2021.05.075.
110. Lavinia D, Diana M, Ruxandra U, Cristian O. Noninvasive ventilation in neuromuscular diseases. In: Mayank V, ed. *Noninvasive Ventilation in Medicine*. Rijeka: IntechOpen; 2018: Ch. 3.
111. Pinto S, de Carvalho M. Comparison of slow and forced vital capacities on ability to predict survival in ALS. *Amyotroph Lateral Scler Frontotemporal Degener*. 2017;18(7–8):528–533. doi: 10.1080/21678421.2017.1354995.
112. Katz S, Arish N, Rokach A, Zaltzman Y, Marcus EL. The effect of body position on pulmonary function: a systematic review. *BMC Pulm Med*. 2018;18(1):159. doi: 10.1186/s12890-018-0723-4.
113. Ambrosino N, Carpene N, Gherardi M. Chronic respiratory care for neuromuscular diseases in adults. *Eur Respir J*. 2009;34(2):444–451. doi: 10.1183/09031936.00182208.
114. Bach JR, Campagnolo DI, Hoeman S. Life satisfaction of individuals with Duchenne muscular dystrophy using long-term mechanical ventilatory support. *Am J Phys Med Rehabil*. 1991;70(3):129–135. doi: 10.1097/00002060-199106000-00004.
115. Sancho J, Martinez D, Bures E, Diaz JL, Ponz A, Servera E. Bulbar impairment score and survival of stable amyotrophic lateral sclerosis patients after noninvasive ventilation initiation. *ERJ Open Res*. 2018;4(2). doi:10.1183/23120541.00159-2017.
116. Fasano A, Fini N, Ferraro D, Ferri L, Vinceti M, Mandrioli J. Percutaneous endoscopic gastrostomy, body weight loss and survival in amyotrophic lateral sclerosis: a population-based registry study. *Amyotroph Lateral Scler Frontotemporal Degener*. 2017;18(3–4):233–242. doi: 10.1080/21678421.2016.1270325.

117. Strijbos D, Hofstede J, Keszthelyi D, Masclee AAM, Gilissen LPL. Percutaneous endoscopic gastrostomy under conscious sedation in patients with amyotrophic lateral sclerosis is safe: an observational study. *Eur J Gastroenterol Hepatol*. 2017;29(11):1303–1308. doi: 10.1097/MEG.0000000000000959.
118. Park JH, Kang SW. Percutaneous radiologic gastrostomy in patients with amyotrophic lateral sclerosis on non-invasive ventilation. *Arch Phys Med Rehabil*. 2009;90(6):1026–1029. doi: 10.1016/j.apmr.2008.12.006.
119. Gaspar R, Ramalho R, Coelho R, Andrade P, Goncalves MR, Macedo G. Percutaneous endoscopic gastrostomy placement under NIV in amyotrophic lateral sclerosis with severe ventilatory dysfunction: a safe and effective procedure. *GE Port J Gastroenterol*. 2023;30(1):61–67. doi: 10.1159/000519926.
120. Tzeng AC, Bach JR. Prevention of pulmonary morbidity for patients with neuromuscular disease. *Chest*. 2000;118 (5):1390–1396. doi: 10.1378/chest.118.5.1390.
121. Chatwin M, Toussaint M, Gonçalves MR, et al. Airway clearance techniques in neuromuscular disorders: a state of the art review. *Respir Med*. 2018;136:98–110. doi: 10.1016/j.rmed.2018.01.012.
122. Chatwin M, Simonds AK. Long-term mechanical insufflation-exsufflation cough assistance in neuromuscular disease: patterns of use and lessons for application. *Respir Care*. 2020;65(2):135–143. doi: 10.4187/respcare.06882.
123. Heritier Barras AC, Adler D, Iancu Ferfoglia R, et al. Is tracheostomy still an option in amyotrophic lateral sclerosis? Reflections of a multidisciplinary work group. *Swiss Med Wkly*. 2013;143(3132):w13830. doi: 10.4414/smw.2013.13830.
124. Castro-Añón O, Pérez de Llano LA, De la Fuente Sánchez S, et al. Obesity-hypoventilation syndrome: increased risk of death over sleep apnea syndrome. *PLOS ONE*. 2015;10(2):e0117808. doi: 10.1371/journal.pone.0117808.
125. Borel JC, Borel AL, Monneret D, Tamisier R, Levy P, Pepin JL. Obesity hypoventilation syndrome: from sleep-disordered breathing to systemic comorbidities and the need to offer combined treatment strategies. *Respirology*. 2012;17(4):601–610. doi: 10.1111/j.1440-1843.2011.02106.x.
126. Randerath W, Verbraecken J, Andreas S, et al. Definition, discrimination, diagnosis and treatment of central breathing disturbances during sleep. *Eur Respir J*. 2017;49(1):1600959. doi: 10.1183/13993003.00959-2016.
127. Kaw R, Kaminska M. Obesity hypoventilation: traditional versus nontraditional populations. *Sleep Med Clin*. 2020;15 (4):449–459. doi: 10.1016/j.jsmc.2020.08.001.
128. Mokhlesi B, Masa JF, Brozek JL, et al. Evaluation and management of obesity hypoventilation syndrome. An official American Thoracic Society clinical practice guideline. *Am J Respir Crit Care Med*. 2019;200(3):e6–e24. doi: 10.1164/rccm.201905-1071ST.
129. Masa JF, Pépin JL, Borel JC, Mokhlesi B, Murphy PB, Sánchez-Quiroga M. Obesity hypoventilation syndrome. *Eur Respir Rev*. 2019;28(151):180097. doi: 10.1183/16000617.0097-2018.
130. Le KDR, Le K, Foo F. The impact of glucagon-like peptide 1 receptor agonists on obstructive sleep apnoea: a scoping review. *Pharmacy (Basel)*. 2024;12(1):11. doi: 10.3390/pharmacy12010011.
131. Masa JF, Mokhlesi B, Benítez I, et al. Cost-effectiveness of positive airway pressure modalities in obesity hypoventilation syndrome with severe obstructive sleep apnoea. *Thorax*. 2020;75(6):459–467. doi: 10.1136/thoraxjnl-2019-213622.
132. Murphy PB, Patout M, Arbane G, et al. Cost-effectiveness of outpatient versus inpatient non-invasive ventilation setup in obesity hypoventilation syndrome: the OPIP trial. *Thorax*. 2023;78(1):24–31. doi: 10.1136/thorax-2021-218497.
133. Masa JF, Corral J, Alonso ML, et al. Efficacy of different treatment alternatives for obesity hypoventilation syndrome. Pickwick study. *Am J Respir Crit Care Med*. 2015;192(1):86–95. doi: 10.1164/rccm.201410-1900OC.
134. Piper AJ, Wang D, Yee BJ, Barnes DJ, Grunstein RR. Randomised trial of CPAP vs bilevel support in the treatment of obesity hypoventilation syndrome without severe nocturnal desaturation. *Thorax*. 2008;63(5):395–401. doi: 10.1136/thx.2007.081315.
135. Howard ME, Piper AJ, Stevens B, et al. A randomised controlled trial of CPAP versus non-invasive ventilation for initial treatment of obesity hypoventilation syndrome. *Thorax*. 2017;72(5):437–444. doi: 10.1136/thoraxjnl-2016-208559.
136. Masa JF, Mokhlesi B, Benítez I, et al. Long-term clinical effectiveness of continuous positive airway pressure therapy versus non-invasive ventilation therapy in patients with obesity hypoventilation syndrome: a multicentre, open-label, randomised controlled trial. *Lancet*. 2019;393(10182):1721–1732. doi: 10.1016/S0140-6736(18)32978-7.
137. Arellano-Maric MP, Hamm C, Duiverman ML, et al. Obesity hypoventilation syndrome treated with non-invasive ventilation: is a switch to CPAP therapy feasible? *Respirology*. 2020;25(4):435–442. doi: 10.1111/resp.13704.
138. Corral J, Mogollon MV, Sánchez-Quiroga M, et al. Echocardiographic changes with non-invasive ventilation and CPAP in obesity hypoventilation syndrome. *Thorax*. 2018;73(4):361–368. doi: 10.1136/thoraxjnl-2017-210642.
139. Neborak JM, Nowalk NC, Mokhlesi B. The overlap of obesity-hypoventilation syndrome and obstructive sleep apnea: how to treat? *Arch Bronconeumol*. 2022;58(7):531–532. doi: 10.1016/j.arbres.2021.08.012.
140. de Llano LP, Castro-Añón O, Castro-Cabana L, Méndez Marote L, Golpe R. Long-term effectiveness of CPAP in patients with severe obesity-hypoventilation syndrome. *Sleep Breath*. 2021;25(2):947–950.
141. Georgoulis M, Yiannakouris N, Kechribari I, et al. Dose-response relationship between weight loss and improvements in obstructive sleep apnea severity after a diet/lifestyle interventions: secondary analyses of the "mimosa" randomized clinical trial. *J Clin Sleep Med*. 2022;18(5):1251–1261. doi: 10.5664/jcsm.9834.

142. Shneerson JM, Simonds AK. Noninvasive ventilation for chest wall and neuromuscular disorders. *Eur Respir J*. 2002;20(2):480–487. doi: 10.1183/09031936.02.00404002.
143. McKim DA, Road J, Avendano M, et al. Home mechanical ventilation: a Canadian Thoracic Society clinical practice guideline. *Can Respir J*. 2011;18(4):197–215. doi: 10.1155/2011/139769.
144. Martí S, Pallero M, Ferrer J, et al. Predictors of mortality in chest wall disease treated with noninvasive home mechanical ventilation. *Respir Med*. 2010;104(12):1843–1849. doi: 10.1016/j.rmed.2010.08.013.
145. Duiverman ML, Bladder G, Meinesz AF, Wijkstra PJ. Home mechanical ventilatory support in patients with restrictive ventilatory disorders: a 48-year experience. *Respir Med*. 2006;100(1):56–65. doi: 10.1016/j.rmed.2005.04.015.
146. Pierucci P, Crimi C, Carlucci A, et al. Reinvent: ERS international survey on restrictive thoracic diseases in long term home noninvasive ventilation. *ERJ Open Res*. 2021;7(2):00911–2020. doi: 10.1183/23120541.00911-2020.
147. Nauffal D, Doménech R, Martínez García MA, Compte L, Macián V, Perpiñá M. Noninvasive positive pressure home ventilation in restrictive disorders: outcome and impact on health-related quality of life. *Respir Med*. 2002;96(10):777–783. doi: 10.1053/rmed.2002.1347.
148. Tuggey JM, Elliott MW. Randomised crossover study of pressure and volume non-invasive ventilation in chest wall deformity. *Thorax*. 2005;60(10):859–864. doi: 10.1136/thx.2005.041327.
149. Nicolini A, Barlascini C, Piroddi IM, Garuti G, Banfi PI. Effectiveness and safety of mouthpiece ventilation and nocturnal non-invasive ventilation in patients with kyphoscoliosis: short and long-term outcomes after an episode of acute respiratory failure. *Rev Port Pneumol (2006)*. 2016;22(2):75–81. doi: 10.1016/j.rppnen.2015.09.009.
150. Durao V, Grafino M, Pamplona P. Chronic respiratory failure in patients with chronic obstructive pulmonary disease under home noninvasive ventilation: real-life study. *Pulmonology*. 2018;24(5):280–288. doi: 10.1016/j.pulmoe.2018.02.007.
151. Kotanen P, Brander P, Kreivi HR. The prevalence of non-invasive ventilation and long-term oxygen treatment in Helsinki University Hospital area, Finland. *BMC Pulm Med*. 2022;22(1):248. doi: 10.1186/s12890-022-02044-5.
152. Macrea M, Oczkowski S, Rochwerg B, et al. Long-term noninvasive ventilation in chronic stable hypercapnic chronic obstructive pulmonary disease. An official American Thoracic Society clinical practice guideline. *Am J Respir Crit Care Med*. 2020;202(4):e74–e87. doi: 10.1164/rccm.202006-2382ST.
153. Chiu HY, Chen PY, Chuang LP, et al. Diagnostic accuracy of the Berlin questionnaire, STOP-BANG, STOP, and Epworth sleepiness scale in detecting obstructive sleep apnea: a bivariate meta-analysis. *Sleep Med Rev*. 2017;36:57–70. doi: 10.1016/j.smrv.2016.10.004.
154. Oliveira MG, Nery LE, Santos-Silva R, et al. Is portable monitoring accurate in the diagnosis of obstructive sleep apnea syndrome in chronic pulmonary obstructive disease? *Sleep Med*. 2012;13(8):1033–1038. doi: 10.1016/j.sleep.2012.06.011.
155. Chang Y, Xu L, Han F, et al. Validation of the Nox-T3 portable monitor for diagnosis of obstructive sleep apnea in patients with chronic obstructive pulmonary disease. *J Clin Sleep Med*. 2019;15(4):587–596. doi: 10.5664/jcsm.7720.
156. Struik FM, Sprooten RT, Kerstjens HA, et al. Nocturnal non-invasive ventilation in COPD patients with prolonged hypercapnia after ventilatory support for acute respiratory failure: a randomised, controlled, parallel-group study. *Thorax*. 2014;69(9):826–834. doi: 10.1136/thoraxjnl-2014-205126.
157. Murphy PB, Rehal S, Arbane G, et al. Effect of home noninvasive ventilation with oxygen therapy vs oxygen therapy alone on hospital readmission or death after an acute COPD exacerbation: a randomized clinical trial. *Jama*. 2017;317(21):2177–2186. doi: 10.1001/jama.2017.4451.
158. Suraj KP, Jyothi E, Rakhi R. Role of domiciliary noninvasive ventilation in chronic obstructive pulmonary disease patients requiring repeated admissions with acute type II respiratory failure: a prospective cohort study. *Indian J Crit Care Med*. 2018;22(6):397–401. doi: 10.4103/ijccm.IJCCM_61_18.
159. Tuggey JM, Plant PK, Elliott MW. Domiciliary non-invasive ventilation for recurrent acidotic exacerbations of COPD: an economic analysis. *Thorax*. 2003;58(10):867–871.
160. Galli JA, Krahnke JS, James Mamary A, Shenoy K, Zhao H, Criner GJ. Home non-invasive ventilation use following acute hypercapnic respiratory failure in COPD. *Respir Med*. 2014;108(5):722–728. doi: 10.1016/j.rmed.2014.03.006.
161. Struik FM, Lacasse Y, Goldstein RS, Kerstjens HA, Wijkstra PJ. Nocturnal noninvasive positive pressure ventilation in stable COPD: a systematic review and individual patient data meta-analysis. *Respir Med*. 2014;108(2):329–337. doi: 10.1016/j.rmed.2013.10.007.
162. Oscroft NS, Chadwick R, Davies MG, Quinnell TG, Smith IE. Volume assured versus pressure preset non-invasive ventilation for compensated ventilatory failure in COPD. *Respir Med*. 2014;108(10):1508–1515. doi: 10.1016/j.rmed.2014.07.010.
163. Dreher M, Storre JH, Schmoor C, Windisch W. High-intensity versus low-intensity non-invasive ventilation in patients with stable hypercapnic COPD: a randomised crossover trial. *Thorax*. 2010;65(4):303–308. doi: 10.1136/thx.2009.124263.
164. Murphy PB, Brignall K, Moxham J, Polkey MI, Davidson AC, Hart N. High pressure versus high intensity noninvasive ventilation in stable hypercapnic chronic obstructive pulmonary disease: a randomized crossover trial. *Int J Chron Obstruct Pulmon Dis*. 2012;7:811–818.

165. Windisch W, Haenel M, Storre JH, Dreher M. High-intensity non-invasive positive pressure ventilation for stable hypercapnic COPD. *Int J Med Sci*. 2009;6(2):72–76. doi: 10.7150/ijms.6.72.
166. Marrara KT, Di Lorenzo VAP, Jaenisch RB, et al. Noninvasive ventilation as an important adjunct to an exercise training program in subjects with moderate to severe COPD. *Respir Care*. 2018;63(11):1388–1398. doi: 10.4187/respcare.05763.
167. Xiang G, Wu Q, Wu X, Hao S, Xie L, Li S. Non-invasive ventilation intervention during exercise training in individuals with chronic obstructive pulmonary disease: a systematic review and meta-analysis. *Ann Phys Rehabil Med*. 2021;64(6):101460. doi: 10.1016/j.rehab.2020.101460.
168. Archangelidi O, Carr SB, Simmonds NJ, Bilton D, Banya W, Cullinan P. Non-invasive ventilation and clinical outcomes in cystic fibrosis: findings from the UK CF registry. *J Cyst Fibros*. 2019;18(5):665–670. doi: 10.1016/j.jcf.2018.11.006.
169. Fauroux B, Burgel PR, Boelle PY, et al. Practice of noninvasive ventilation for cystic fibrosis: a nationwide survey in France. *Respir Care*. 2008;53(11):1482–1489.
170. Wadsworth LE, Belcher J, Bright-Thomas RJ. Non-invasive ventilation is associated with long-term improvements in lung function and gas exchange in cystic fibrosis adults with hypercapnic respiratory failure. *J Cyst Fibros*. 2021;20(5):e40–e5.
171. Flight WG, Shaw J, Johnson S, et al. Long-term non-invasive ventilation in cystic fibrosis – experience over two decades. *J Cyst Fibros*. 2012;11(3):187–192. doi: 10.1016/j.jcf.2011.11.006.
172. Wedzicha JA, Muir JF. Noninvasive ventilation in chronic obstructive pulmonary disease, bronchiectasis and cystic fibrosis. *Eur Respir J*. 2002;20(3):777–784. doi: 10.1183/09031936.02.00308502.
173. Young AC, Wilson JW, Kotsimbos TC, Naughton MT. Randomised placebo controlled trial of non-invasive ventilation for hypercapnia in cystic fibrosis. *Thorax*. 2008;63(1):72–77. doi: 10.1136/thx.2007.082602.
174. Serra A, Polese G, Braggion C, Rossi A. Non-invasive proportional assist and pressure support ventilation in patients with cystic fibrosis and chronic respiratory failure. *Thorax*. 2002;57(1):50–54.
175. Hodson ME, Madden BP, Steven MH, Tsang VT, Yacoub MH. Non-invasive mechanical ventilation for cystic fibrosis patients–a potential bridge to transplantation. *Eur Respir J*. 1991;4(5):524–527. doi: 10.1183/09031936.93.04050524.
176. Morrell MR, Kiel SC, Pilewski JM. Organ transplantation for cystic fibrosis. *Semin Respir Crit Care Med*. 2019;40(6):842–856. doi: 10.1055/s-0039-3399554.
177. Moran F, Bradley JM, Piper AJ. Non-invasive ventilation for cystic fibrosis. *Cochrane Database Syst Rev*. 2017;2(2):Cd002769.
178. Stanford G, Parrott H, Bilton D, Agent P. Positive pressure–analysing the effect of the addition of non-invasive ventilation (NIV) to home airway clearance techniques (ACT) in adult cystic fibrosis (CF) patients. *Physiother Theory Pract*. 2015;31(4):270–274. doi: 10.3109/09593985.2014.994151.
179. Soltaninejad F, Samim A, Salmasi M, et al. Non invasive ventilation's effectiveness (NIV) in patients with interstitial lung disease and hypercapnic respiratory failure. *Tanaffos*. 2022;21(3):302–306.
180. Cuerpo S, Palomo M, Hernández-González F, et al. Improving home oxygen therapy in patients with interstitial lung diseases: application of a noninvasive ventilation device. *Ther Adv Respir Dis*. 2020;14:1753466620963027. doi: 10.1177/1753466620963027.
181. Moderno EV, Yamaguti WP, Schettino GP, et al. Effects of proportional assisted ventilation on exercise performance in idiopathic pulmonary fibrosis patients. *Respir Med*. 2010;104(1):134–141. doi: 10.1016/j.rmed.2009.08.001.
182. Bornitz F, Ewert R, Knaak C, Magnet FS, Windisch W, Herth F. Weaning from invasive ventilation in specialist centers following primary weaning failure. *Dtsch Arztebl International*. 2020;117(12):205–210. doi: 10.3238/arztebl.2020.0205.
183. Dreyer P, Lorenzen CK, Schou L, Felding M. Survival in ALS with home mechanical ventilation non-invasively and invasively: a 15-year cohort study in West Denmark. *Amyotroph Lateral Scler Frontotemporal Degener*. 2014;15(1–2):62–67. doi: 10.3109/21678421.2013.837929.
184. Spittel S, Maier A, Kettemann D, et al. Non-invasive and tracheostomy invasive ventilation in amyotrophic lateral sclerosis: utilization and survival rates in a cohort study over 12 years in Germany. *Eur J Neurol*. 2021;28(4):1160–1171. doi: 10.1111/ene.14647.
185. Huttmann SE, Magnet FS, Karagiannidis C, Storre JH, Windisch W. Quality of life and life satisfaction are severely impaired in patients with long-term invasive ventilation following ICU treatment and unsuccessful weaning. *Ann Intensive Care*. 2018;8(1):38. doi: 10.1186/s13613-018-0384-8.
186. Schwarz EI, Bloch KE. Frontiers in clinical practice of long-term care of chronic ventilatory failure. *Respiration*. 2019;98(1):1–15. doi: 10.1159/000499316.
187. Kotanen P, Kreivi HR, Vainionpää A, Laaksovirta H, Brander P, Siirala W. Home invasive mechanical ventilation in Finland in 2015-2019. *ERJ Open Res*. 2020;6(4):00223–2020. doi: 10.1183/23120541.00223-2020.
188. Winck JC. Long-term tracheostomy ventilation in Portugal: survey based on home care providers. *Pulmonology*. 2019;25(3):180. doi: 10.1016/j.pulmoe.2019.01.006.
189. Mifsud Bonnici D, Sanctuary T, Warren A, et al. Prospective observational cohort study of patients with weaning failure admitted to a specialist weaning, rehabilitation and home mechanical ventilation centre. *BMJ Open*. 2016;6(3):e010025. doi: 10.1136/bmjopen-2015-010025.

190. Heine A, Hustig-Kittler V, Schröder M, et al. Evaluation of weaning success in long-term invasively ventilated patients at a weaning center. *Pneumologie*. 2021;75(6):447–452. doi: 10.1055/a-1346-6208.
191. Withers A, Ching Man TC, D'Cruz R, et al. Highlights from the Respiratory Failure and Mechanical Ventilation 2020 conference. *ERJ Open Res*. 2021;7(1):00752–2020. doi: 10.1183/23120541.00752-2020.
192. Brown J. Tracheostomy to noninvasive ventilation: from acute care to home. *Sleep Med Clinic*. 2020;15(4):593–598. doi: 10.1016/j.jsmc.2020.08.003.
193. Simonds AK. Home mechanical ventilation: an overview. *Ann Am Thorac Soc*. 2016;13(11):2035–2044. doi: 10.1513/AnnalsATS.201606-454FR.
194. Dyrstad DN, Hansen BS, Gundersen EM. Factors that influence user satisfaction: tracheotomised home mechanical ventilation users' experiences. *J Clin Nurs*. 2013;22(3–4):331–338. doi: 10.1111/j.1365-2702.2012.04304.x.
195. Helsedirektoratet. Nasjonal faglig retningslinje for langtids mekanisk ventilasjon (LTMV) (national professional guideline for long-term mechanical ventilation). 2012. https://www.helsedirektoratet.no/retningslinjer/langtids-mekanisk-ventilasjon-ltmv/Langtids%20mekanisk%20ventilasjon%20(LTMV)%20%E2%80%93%20Nasjonal%20faglig%20retningslinje.pdf/_/attachment/inline/e8acb43e-3d88-45e8-958b-561137699474:4589070adf4ca8bbf2f7ce57907c3818887f0e26/Langtids%20mekanisk%20ventilasjon%20(LTMV)%20%E2%80%93%20Nasjonal%20faglig%20retningslinje.pdf.
196. Kaub-Wittemer D, Steinbüchel N, Wasner M, Laier-Groeneveld G, Borasio GD. Quality of life and psychosocial issues in ventilated patients with amyotrophic lateral sclerosis and their caregivers. *J Pain Symptom Manage*. 2003;26(4):890–896. doi: 10.1016/S0885-3924(03)00323-3.
197. Sutt AL, Cornwell P, Mullany D, Kinneally T, Fraser JF. The use of tracheostomy speaking valves in mechanically ventilated patients results in improved communication and does not prolong ventilation time in cardiothoracic intensive care unit patients. *J Crit Care*. 2015;30(3):491–494. doi: 10.1016/j.jcrc.2014.12.017.
198. Zaga CJ, Berney S, Hepworth G, et al. Tracheostomy clinical practices and patient outcomes in three tertiary metropolitan hospitals in Australia. *Aust Crit Care*. 2023;36(3):327–335. doi: 10.1016/j.aucc.2022.03.002.
199. St-Laurent A, Zielinski D, Qazi A, et al. Chronic tracheostomy care of ventilator-dependent and -independent children: clinical practice patterns of pediatric respirologists in a publicly funded (Canadian) healthcare system. *Pediatr Pulmonol*. 2023;58(1):140–151. doi: 10.1002/ppul.26171.
200. Fried-Oken M, Mooney A, Peters B. Supporting communication for patients with neurodegenerative disease. *NeuroRehabilitation*. 2015;37(1):69–87. doi: 10.3233/NRE-151241.
201. Nakayama Y, Shimizu T, Mochizuki Y, et al. Predictors of impaired communication in amyotrophic lateral sclerosis patients with tracheostomy-invasive ventilation. *Amyotroph Lateral Scler Frontotemporal Degener*. 2015;17(1–-2):38–46. doi: 10.3109/21678421.2015.1055276.
202. Khalaila R, Zbidat W, Anwar K, Bayya A, Linton DM, Sviri S. Communication difficulties and psychoemotional distress in patients receiving mechanical ventilation. *Am J Critical Care*. 2011;20(6):470–479. doi: 10.4037/ajcc2011989.
203. Radbruch L, De Lima L, Knaul F, et al. Redefining palliative care-a new consensus-based definition. *J Pain Symptom Manage*. 2020;60(4):754–764. doi: 10.1016/j.jpainsymman.2020.04.027.
204. Lei n.º 52/2012 de 5 de setembro - Lei de Bases dos Cuidados Paliativos. Diário da República, 1.a série N.o 172 de 5 de setembro de 2012: 5119-5124. https://files.diariodarepublica.pt/1s/2012/09/17200/0511905124.pdf. Accessed March 1, 2023.
205. Marsaa K, Gundestrup S, Jensen JU, et al. Danish respiratory society position paper: palliative care in patients with chronic progressive non-malignant lung diseases. *Eur Clin Respir J*. 2018;5(1):1530029. doi: 10.1080/20018525.2018.1530029.
206. Lanken PN, Terry PB, Delisser HM, et al. An official American Thoracic Society clinical policy statement: palliative care for patients with respiratory diseases and critical illnesses. *Am J Respir Crit Care Med*. 2008;177(8):912–927. doi: 10.1164/rccm.200605-587ST.
207. Sullivan DR, Iyer AS, Enguidanos S, et al. Palliative care early in the care continuum among patients with serious respiratory illness: an official ATS/AAHPM/HPNA/SWHPN policy statement. *Am J Respir Crit Care Med*. 2022;206(6):e44–e69. doi: 10.1164/rccm.202207-1262ST.
208. Entidade Reguladora da Saúde. Direitos e Deveres dos Utentes dos Serviços de Saúde. https://www.ers.pt/media/pqll00ec/direitos-e-deveres-dos-utentes-dos-servi%C3%A7os-de-sa%C3%BAde.pdf.
209. Pereira RB, Joana D, Drummond M. *Ventilaçao Não Invasiva Na Esclerose Lateral Amiotrófica: Adaptaçao Em contexto agudo Vs programado e a sua correlação prognóstica*. Porto: II Congresso de Cuidados Respiratórios; 2024.
210. Rietjens JAC, Sudore RL, Connolly M, et al. Definition and recommendations for advance care planning: an international consensus supported by the European Association for Palliative Care. *Lancet Oncol*. 2017;18(9):e543–e551. doi: 10.1016/S1470-2045(17)30582-X.
211. Ribeiro C, Pamplona P, Simonds AK. Advance care planning in patients with respiratory failure. *Euro Respir Rev*. 2024;33(174):240120. doi: 10.1183/16000617.0120-2024.
212. Dingfield LE, Kayser JB. Integrating advance care planning into practice. *Chest*. 2017;151(6):1387–1393. doi: 10.1016/j.chest.2017.02.024.

213. Payne S, Harding A, Williams T, Ling J, Ostgathe C. Revised recommendations on standards and norms for palliative care in Europe from the European Association for Palliative Care (EAPC): a Delphi study. *Palliat Med*. 2022;36 (4):680–697. doi: 10.1177/02692163221074547.
214. Sudore RL, Lum HD, You JJ, et al. Defining advance care planning for adults: a consensus definition from a multidisciplinary Delphi panel. *J Pain Symptom Manage*. 2017;53(5):821e1–832e1. doi: 10.1016/j.jpainsymman.2016.12.331.
215. Lei n.º 25/2012 de 16 de julho - Regula as diretivas antecipadas de vontade, designadamente sob a forma de testamento vital, e a nomeação de procurador de cuidados de saúde e cria o Registo Nacional do Testamento Vital (RENTEV). Dr 1ª série 136: 3728-3730. https://www.pgdlisboa.pt/leis/lei_mostra_articulado.php?nid=1765&tabela=leis&so_miolo=. Accessed May 1, 2023.
216. Soriani MH, Desnuelle C. Care management in amyotrophic lateral sclerosis. *Rev Neurol (Paris)*. 2017;173 (5):288–299. doi: 10.1016/j.neurol.2017.03.031.
217. Simonds AK. Recent advances in respiratory care for neuromuscular disease. *Chest*. 2006;130(6):1879–1886. doi: 10.1378/chest.130.6.1879.
218. Van Damme P, Al-Chalabi A, Andersen PM, et al. European Academy of Neurology (EAN) guideline on the management of amyotrophic lateral sclerosis in collaboration with European Reference Network for Neuromuscular Diseases (ERN EURO-NMD). *Eur J Neurol*. 2024;31(6):e16264. doi: 10.1111/ene.16264.
219. Bausewein C, Currow DC, Johnson MJ, eds. *Palliative Care in Respiratory Disease*. Lausanne, Switzerland: European Respiratory Society (ERS). 2016: 258. doi: 10.1183/2312508X.erm7316
220. Spurr L. The treatment burden of long-term home noninvasive ventilation. *Breathe (Sheff)*. 2021;17(1):200291. doi: 10.1183/20734735.0291-2020.
221. Curtis JR, Cook DJ, Sinuff T, et al. Noninvasive positive pressure ventilation in critical and palliative care settings: understanding the goals of therapy. *Crit Care Med*. 2007;35(3):932–939. doi: 10.1097/01.CCM.0000256725.73993.74.
222. Cortegiani A, Mercadante S, Gregoretti C. Palliative ventilatory support: same knowledge, different goal. *J Thorac Dis*. 2018;10(3):E236–e7.
223. Nava S, Ferrer M, Esquinas A, et al. Palliative use of non-invasive ventilation in end-of-life patients with solid tumours: a randomised feasibility trial. *Lancet Oncol*. 2013;14(3):219–227. doi: 10.1016/S1470-2045(13)70009-3.
224. Carlucci A, Guerrieri A, Nava S. Palliative care in COPD patients: is it only an end-of-life issue? *Eur Respir Rev*. 2012;21 (126):347–354. doi: 10.1183/09059180.00001512.
225. Bausewein C, Daveson BA, Currow DC, et al. EAPC White paper on outcome measurement in palliative care: improving practice, attaining outcomes and delivering quality services – recommendations from the European association for palliative care (EAPC) task force on outcome measurement. *Palliat Med*. 2016;30(1):6–22. doi: 10.1177/0269216315589898.
226. Crimi C, Pierucci P, Carlucci A, Cortegiani A, Gregoretti C. Long-term ventilation in neuromuscular patients: review of concerns, beliefs, and ethical dilemmas. *Respiration*. 2019;97(3):185–196. doi: 10.1159/000495941.
227. Oliver DJ. Palliative care in motor neurone disease: where are we now? *Palliat Care*. 2019;12:1178224218813914. doi: 10.1177/1178224218813914.
228. Oliver DJ. Palliative care for patients with motor neurone disease: current challenges. *Degener Neurol Neuromuscul Dis*. 2016;6:65–72. doi: 10.2147/DNND.S85103.
229. Lei n.º 31/2018 - Direitos das pessoas em contexto de doença avançada e em fim de vida. Diário da República, 1.a série—n.o 137—18 de julho de 2018:3238-3239. https://www.pgdlisboa.pt/leis/lei_mostra_articulado.php?nid=2917&tabela=leis&ficha=1&pagina=1&so_miolo=. Accessed May 1, 2023.
230. Escarrabill J, Vianello A, Farrero E, Ambrosino N, Martínez Llorens J, Vitacca M. Place of death in patients with amyotrophic lateral sclerosis. *Rev Port Pneumol*. 2014;20(4):188–193. doi: 10.1016/j.rppneu.2014.03.001.
231. Gomes B, Sarmento VP, Ferreira PL, Higginson IJ. Epidemiological study of place of death in Portugal in 2010 and comparison with the preferences of the Portuguese population. *Acta Med Port*. 2013;26(4):327–334. doi: 10.20344/amp.429.
232. Decreto Regulamentar n.º 1/2022 de 10 de janeiro - estatuto de cuidador informal. https://eportugal.gov.pt/guias/cuidador-informal/descanso-do-cuidador/. Accessed October 1, 2023.
233. Farquhar M. Supporting informal carers. *ERS Monogr*. 2016;73:51–69. doi: 10.1183/2312508X.10011315.
234. Baxter SK, Baird WO, Thompson S, et al. The use of non-invasive ventilation at end of life in patients with motor neurone disease: a qualitative exploration of family carer and health professional experiences. *Palliat Med*. 2013;27 (6):516–523. doi: 10.1177/0269216313478449.
235. Cullum KG, Madani CA, Cutler ER, Reavis KJ. The lived experience of respiratory therapists during withdrawal of advanced life-sustaining therapies at end of life in the ICU. *Respir Care*. 2022;67(12):1568–1577. doi: 10.4187/respcare.09249.
236. Regulamento n.º 707/2016, de 21 de Julho - Regulamento de Deontologia Médica. https://www.pgdlisboa.pt/leis/lei_mostra_articulado.php?artigo_id=2649A0067&nid=2649&tabela=leis&pagina=1&ficha=1&so_miolo=&nversao=#artigo). Accessed October 1, 2023.
237. Phelps K, Regen E, Oliver D, McDermott C, Faull C. Withdrawal of ventilation at the patient's request in MND: a retrospective exploration of the ethical and legal issues that have arisen for doctors in the UK. *BMJ Support Palliat Care*. 2017;7(2):189–196. doi: 10.1136/bmjspcare-2014-000826.

238. Ireland AfPMoGBa. *Withdrawal of Ventilation at the Request of a Patient with Motor Neurone Disease: Guidance for Professionals*. Association for Palliative Medicine of Great Britain and Ireland. 2015. https://apmonline.org/wp-content/uploads/Guidance-Final-November-2024-v2.pdf. Accessed March 2024.
239. Lei n.º 22/2023 - Regula as condições em que a morte medicamente assistida não é punível e altera o Código Penal. Diário da República, 1.a série n.o 101 de 25 de maio de 2023:10-20. https://www.pgdlisboa.pt/leis/lei_mostra_articulado.php?artigo_id=3648A0027&nid=3648&tabela=leis&pagina=1&ficha=1&so_miolo=&nversao=. Accessed September 1, 2023.
240. Goldstein NE, Cohen LM, Arnold RM, Goy E, Arons S, Ganzini L. Prevalence of formal accusations of murder and euthanasia against physicians. *J Palliat Med*. 2012;15(3):334–339. doi: 10.1089/jpm.2011.0234.
241. Lundorff M, Holmgren H, Zachariae R, Farver-Vestergaard I, O'Connor M. Prevalence of prolonged grief disorder in adult bereavement: a systematic review and meta-analysis. *J Affect Disord*. 2017;212:138–149. doi: 10.1016/j.jad.2017.01.030.
242. DGS NORMA 003/2019 Modelo de Intervenção Diferenciada no Luto Prolongado em Adultos. https://normas.dgs.min-saude.pt/2019/04/23/modelo-de-intervencao-diferenciada-no-luto-prolongado-em-adultos/. Accessed October 1, 2023.