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Chronic Neuromuscular Respiratory Failure and Home Assisted Ventilation

Hugo Carmona,[1] Andrew D. Graustein,[1,2] and Joshua O. Benditt[1]

[1]Division of Pulmonary, Critical Care and Sleep Medicine, University of Washington School of Medicine, Seattle, Washington, USA; email: hcarmona@uw.edu, benditt@uw.edu

[2]VA Puget Sound Health Care System, Seattle, Washington, USA; email: andrew.graustein@va.gov

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Keywords

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Abstract

Chronic respiratory failure is a common, important complication of many types of neuromuscular and chest wall disorders. While the pathophysiology of each disease may be different, these disorders can variably affect all muscles involved in breathing, including inspiratory, expiratory, and bulbar muscles, ultimately leading to chronic respiratory failure and hypoventilation. The use of home assisted ventilation through noninvasive interfaces aims to improve the symptoms of hypoventilation, improve sleep quality, and, when possible, improve mortality. An increasing variety of interfaces has allowed for improved comfort and compliance. In a minority of scenarios, noninvasive ventilation is either not appropriate or no longer effective due to disease progression, and a transition to tracheal ventilation should be considered.

INTRODUCTION

Chronic respiratory failure is a common, important complication of many types of neuromuscular and chest wall disorders. A patient with otherwise normal lungs can develop respiratory failure through muscular weakness and/or thoracic cage abnormalities. Hypercapnia due to hypoventilation is the predominant issue, although in some patients, hypoxemia due to severe CO_2 elevation or atelectasis can occur. Patients with chronic neuromuscular respiratory failure often need home respiratory support in the form of noninvasive positive pressure ventilation (NPPV) or invasive tracheal ventilation (TV). The use of home ventilation for patients with chronic respiratory failure is variable across geographic locales both in the United States and around the world but is likely rising in frequency (1). Depending on how strictly "ventilator support" is defined, it is estimated that approximately 11,000–48,000 patients in the United States receive some variant of home ventilation, using Medicare claims data from 2010 (2). This is, in part, due to the advances in respiratory care that are allowing children to survive diseases that previously led to death from neuromuscular respiratory failure, such as Duchenne muscular dystrophy (DMD) (3–6), as well as the increasing availability of less expensive NPPV. Despite advances in technology, respiratory failure continues to be the leading cause of morbidity and mortality in patients with neuromuscular disease (NMD) (7), though home ventilation techniques including NPPV and TV have reduced mortality (8, 9) and morbidity, and improved these patients' symptoms and quality of life.

In this review, we briefly cover the pathophysiology of chronic neuromuscular respiratory failure as well as some prototypical diseases that lead to this problem. We then focus on the management of chronic respiratory failure through noninvasive and invasive ventilation, as well as adjunctive therapies to maintain respiratory health in this population.

CHRONIC NEUROMUSCULAR RESPIRATORY FAILURE

Chronic neuromuscular respiratory failure is defined as failure of the inspiratory respiratory muscles (diaphragm, intercostals, and other accessory muscles) to maintain normal ventilation and a normal arterial partial pressure of carbon dioxide ($PaCO_2$). In addition to inspiratory muscle weakness, many patients with NMD also have weakness in the expiratory and bulbar muscles that can impair cough and swallowing and put the patient at even greater risk of major respiratory complications (10).

Pathophysiology

Regardless of etiology, there are common pathways that lead to chronic respiratory failure in NMD (11), as described above. The most common presentation is dyspnea and the effects of increased $PaCO_2$ or low arterial partial pressure of oxygen (PaO_2). Nocturnal hypoventilation is common and is caused by the normal sleep muscular atonia that occurs during rapid eye movement sleep. Sleep atonia affects inspiratory muscles other than the diaphragm (12), but in someone with a weakened diaphragm, this loss of nondiaphragmatic muscle contribution to breathing may be enough to cause nocturnal hypoventilation. Frequent episodes of nocturnal $PaCO_2$ elevation can result in renal compensation with increased bicarbonate ion and, if untreated, can lead to daytime elevation in $PaCO_2$ (11). Symptoms of sleep-disordered breathing, including poor sleep quality, frequent awakening, morning headaches, fatigue, and daytime somnolence should be carefully assessed. Upper airway obstruction due to weak bulbar muscles can also occur, increasing the likelihood of sleep disturbance (13).

Table 1 Examples of nervous system diseases associated with respiratory dysfunction based on pathology location (adapted from Reference 10)

Cortex	Brainstem	Basal ganglia	Spinal cord	Motor nerves	NM junction	Myopathies
Stroke Neoplasm	Stroke Poliomyelitis Multiple sclerosis Multisystem atrophy	Parkinson disease Chorea Dyskinesias	Trauma Transverse myelitis Epidural abscess	Amyotrophic lateral sclerosis Guillain-Barré Vasculitis Critical illness neuropathy	Myasthenia gravis Drugs (NM junction blockers, corticosteroids, anticholinesterase inhibitors) Botulism	Muscular dystrophies Diabetes mellitus Polymyositis

Abbreviation: NM, neuromuscular.

Diseases Leading to Chronic Neuromuscular Respiratory Failure

A variety of disorders can lead to chronic neuromuscular respiratory failure, including neuropathic and myopathic (11) disorders caused by genetic, congenital, and acquired causes. Two major adult populations are afflicted by these diseases: survivors of childhood-onset disorders and those with adult-onset disease.

The prototypical examples of childhood-onset diseases include muscular dystrophies such as DMD, cerebral palsy, and spinal muscle atrophy (4).

A prototypical adult-onset disease is amyotrophic lateral sclerosis (ALS), a progressive motor neuron disease that leads to weakness of all three major respiratory muscle groups: inspiratory, expiratory, and bulbar muscles (14). Other examples of adult-onset chronic respiratory failure include myotonic dystrophy, where respiratory failure does not usually present until the third decade or later (15), and acquired diaphragmatic paralysis such as from critical illness (16). **Table 1** lists some of the NMDs more frequently encountered in clinical practice.

Proper nervous system functioning for the respiratory system requires the cortex of the brain for voluntary breathing, the brainstem for automatic breathing, and the spinal cord and motor neurons, the respiratory muscles, and feedback receptors and nerves for execution (10). Disease at any point in this pathway can lead to respiratory failure. In addition, chest wall diseases (e.g., kyphoscoliosis, flail chest, pectus excavatum) leading to restrictive physiology can contribute to chronic respiratory failure (17).

Clinical Evaluation

A useful way to think about the care of patients with neuromuscular respiratory disease is to break down the neuromuscular system into three areas of function (18, 19):

1. Ventilatory function, which is primarily dependent on inspiratory muscles.
2. Cough function, which is determined by inspiratory and expiratory muscles as well as the muscles controlling the glottis.
3. Swallowing and airway protection, which are determined by the muscles of the glottis.

The clinical evaluation for patients with known and suspected neuromuscular respiratory disease focuses on these three functions of the respiratory system. An initial evaluation should include pulmonary function tests, with special attention to spirometry. The major utility of pulmonary function testing is for staging severity of impairment as well as longitudinal assessment of function over time. The vital capacity can be expected to decrease over time in progressive NMD and can be used to decide when to initiate NPPV (see below). Vital capacity in the supine position can

reveal diaphragm weakness that might otherwise not be obvious. With diaphragm weakness, the supine vital capacity can drop by 20% or more compared to values in the upright posture (20).

Several other measures of neuromuscular strength and function can evaluate ventilatory and cough strength. Maximal inspiratory pressure (MIP) and maximal expiratory pressure (MEP) maneuvers (21) can help track respiratory muscle weakness even in the absence of overall muscle weakness (22), and reduced MIP and MEP are associated with likelihood for hypoventilation and respiratory failure in many conditions, particularly myopathies (23). A weak cough is usually due to a combination of decreasing vital capacity and inspiratory volume, along with weakness of the expiratory muscles (24–26) and sometimes glottic dysfunction (27).

Peak cough flow (PCF) is a widely accepted measure of this vital function, with a reduction in PCF indicating disease severity and increased risk of infection and mortality (28, 29). A peak cough expiratory flow (PCEF) of <270 L/min may be sufficient to clear mucus when a patient is well but puts them at risk of having a low, and thus ineffective, PCEF during respiratory illness (30). Additionally, the longitudinal assessment of the MEP and PCF can help with initiating cough assist maneuvers and devices to help manage secretions and prevent infection (26).

Measurement of CO_2 levels is important in these individuals. Arterial $PaCO_2$ sampling can be difficult and painful for individuals with muscle and blood vessel atrophy. End-tidal ($PetCO_2$) and transcutaneous CO_2 ($PtcCO_2$) monitoring are a more accessible and patient-friendly technology that allows for intermittent and real-time monitoring of daytime and nocturnal hypoventilation (31). As noted above, sleep-disordered breathing is very common in individuals with NMD. In some cases, polysomnography can be helpful to characterize nocturnal hypoventilation; however, it is not essential to initiate nocturnal ventilatory support, as discussed below.

MANAGEMENT OF RESPIRATORY FAILURE BY NONINVASIVE HOME VENTILATION

The use of NPPV is considered standard practice in patients with chronic neuromuscular respiratory failure and can provide long-term ventilatory support of patients with NMD and chest wall disease (32). A recent survey of members of the European Respiratory Society prescribing long-term home NPPV found that, among patients with NMD, ALS was the most common indication (78%), followed by DMD (11%), chest wall deformity (14.5%), and others (4.6%) (33). Below, we discuss which patient populations with chronic respiratory failure are most likely to benefit from NPPV and describe details regarding the timing of NPPV initiation, mask interface options, and ventilatory strategies.

Indications and Goals

The goal of NPPV support is to reduce symptoms of hypoventilation, thereby improving sleep quality and energy, overall quality of life and, if possible, survival. Of the NMDs, ALS has the largest body of evidence supporting the use of NPPV. NPPV prolongs survival in patients with ALS, particularly in those with less severe bulbar dysfunction. This was first described in a retrospective analysis that showed both a survival benefit and a slowing of lung function decline in patients using NPPV for >4 h per day (34). A more recent retrospective analysis suggests a survival benefit associated with extending the use of NPPV to 8 h daily (35). A prospective trial compared outcomes in patients with ALS randomized to either NPPV or standard care when the patients developed orthopnea or symptomatic hypercapnia (36). The use of NPPV prolonged survival overall, but particularly in the subgroup with better bulbar function. Those with severe bulbar dysfunction did not have increased survival, though NPPV did confer some quality-of-life benefits.

A 2017 Cochrane Review noted that, based primarily on this single randomized controlled trial, there is moderate-quality evidence suggesting a survival benefit and low-quality evidence for an improvement in quality of life following use of NPPV in patients with ALS (9). The review notes that future randomized controlled trials are unlikely because not offering NPPV in this patient population would no longer be considered ethical. The review recognizes the importance of future studies examining the timing of NPPV initiation, the benefits of adding cough augmentation, and the health economics of NPPV.

Few prospective studies are available comparing outcomes with and without NPPV in other NMDs and chest well diseases. Retrospective analyses consistently show that survival is prolonged by using NPPV in patients with DMD. Eagle et al. (5) reported mean survival of 19.3 years without NPPV and 25.3 years with NPPV. In patients with DMD in Japan, mean survival was 20.1 years without NPPV and 30.4 years with nasal intermittent positive pressure ventilation (37). The addition of daytime intermittent mouthpiece ventilation to nocturnal nasal intermittent positive pressure ventilation likely prolonged survival and decreased symptom scores in patients with DMD with diurnal hypercapnia (38).

Based on the available evidence, consensus statements exist for the use of NPPV in ALS (39) and DMD (40, 41). A 1998 national consensus conference statement supports the use of NPPV for restrictive lung disease and nocturnal hypoventilation and is based on expert opinion from various American medical societies (42). This statement was reviewed and updated in 2021 (43).

A publication by the Canadian Thoracic Society provides disease-specific practice guidelines for the aforementioned diseases as well as spinal cord injury, postpolio syndrome, kyphoscoliosis, and other causes of chronic respiratory failure (44). It has been noted that while evidence is more limited for the use of NPPV with NMDs other than ALS and DMD, the outcome of untreated hypercapnic respiratory failure is known, raising the question of the appropriateness of randomization to medical therapy rather than NPPV in patients with chronic respiratory failure (18).

Initiation

The decision to initiate NPPV is often based on a combination of patient symptoms and objective findings from pulmonary function tests, arterial blood gases, and overnight polysomnography or desaturation studies. In the United States, limits on reimbursement for NPPV devices may restrict prescribing patterns. The guidelines for initiation of NPPV in NMD and chest wall disorders proposed by various societies and committees are summarized by Hilbert (13). Among respondents to the European Respiratory Society 2019 Assembly survey, the main reason to initiate home NPPV was diurnal hypercapnia, while quality of life/sleep was the most important goal to achieve (33).

However, the optimal timepoint for initiating NPPV is not known. Vital capacity is frequently used to evaluate respiratory function in patients with NMD. In a survey of ALS specialists, practitioners from both the United States and Europe reported that upright forced vital capacity (FVC) was the most common test used to evaluate and trend respiratory function, as well as to decide whether to initiate NPPV (45). A recent retrospective analysis found that very early initiation of NPPV in ALS at FVC $\geq$80% provided a survival advantage (46).

Vital capacity also has prognostic value in patients with ALS, whose FVC at the time of diagnosis is associated with survival (47). A vital capacity threshold is one of the criteria by which patients with restrictive lung disease in the United States can qualify for NPPV device reimbursement from the Centers for Medicare and Medicaid Services (CMS). For patients with NMD or a severe thoracic cage abnormality, CMS reimbursement for NPPV requires the following (48):

1. Documentation of restrictive thoracic disorder.
2. One of the following:
   a. $PaCO_2$ on awake arterial blood gas ≥45 mm Hg while breathing the prescribed fraction of oxygen.
   b. Sleep oximetry with SpO_2 ≤88% for 5 min or more while breathing the prescribed fraction of oxygen.
   c. Either a maximal inspiratory pressure <60 cm H_2O or forced vital capacity <50% predicted.
3. Chronic obstructive pulmonary disease is not a significant contributor to the pulmonary limitation.

However, in the absence of definitive data, there are significant variations in regulatory guidelines for initiating NPPV, which impose financial constraints on the initiation of NPPV. Among surveyed ALS specialists in the United States (45), findings show that, were insurance or other financial constraints not a factor, the majority of ALS specialists would initiate NPPV either with the onset of respiratory symptoms (49.1%) or upon ALS diagnosis (15.8%) rather than at a specific pulmonary function threshold as required by CMS criteria. Only 23% would maintain their current timing of prescribing NPPV. Among European specialists, most ranked the symptoms of orthopnea or dyspnea as the most important reason for prescribing NPPV (45). Notably, current CMS criteria for device reimbursement do not consider respiratory symptoms.

The European Federation of Neurological Societies guidelines recommend initiating NPPV for ALS if any one of the following is present (49, 50):

1. One respiratory clinical symptom related to muscle weakness.
2. FVC <80%.
3. Sniff nasal pressure (SNP) <40 cm H_2O.
4. Significant nocturnal desaturation.
5. pCO_2 >45mm H_2O on a morning arterial blood gas.

While the optimal timing of NPPV initiation for ALS is not known, one common approach would initiate NPPV at the onset of symptoms, and possibly earlier in the disease course should financial constraints allow (50).

Tools other than blood gases, pulse oximetry, and spirometry can be useful for decisions regarding NPPV initiation with NMD. Patients with bulbar dysfunction can have difficulty performing the vital capacity and maximal inspiratory pressure maneuvers due to their inability to form an effective mouth seal around the spirometer or manometer. The SNP, also known as the sniff nasal-inspiratory force (SNIF), can be measured by placing a manometer tube in one nostril while occluding the other nostril. The negative pressure that is generated has prognostic value in patients with ALS (51) and is a component of the American Academy of Neurology's threshold for NPPV consideration (39).

For patients with DMD, an ATS Consensus Statement recommends the use of polysomnography to assess adequacy of home ventilatory support, particularly since nocturnal intermittent positive pressure ventilation usually precedes daytime NPPV (40). The consensus statement does not provide specific criteria for nocturnal NPPV initiation, though it does recommend consideration of daytime NPPV when waking $PaCO_2$ is >50 mm Hg. A DMD care considerations working group recommends the initiation of nocturnal ventilation with one of the following (41):

1. Signs or symptoms of hypoventilation.
2. Baseline SpO_2 <95% with or without $PetCO_2$ >45 mm Hg while awake.

3. An apnea-hypopnea index >10/h, or four or more episodes of desaturation <92%, or desaturation of at least 4% per hour of sleep.

The working group also provides recommendations for initiation of daytime ventilation based on the first two points above. The Canadian Thoracic Society recommends the initiation of NPPV with DMD when there is daytime hypercapnia or symptomatic nocturnal hypoventilation (44).

Interfaces

The interface by which positive pressure is applied to the airways is what differentiates noninvasive from invasive ventilation. Interfaces for NPPV provide ventilation via the nose, the mouth, or a combination of both (32, 52). Nasal interfaces can include a nasal mask covering the entire nose or a nasal pillow in which ventilation is provided via soft plastic prongs into the nares. The oronasal mask covers both the mouth and nose, while a hybrid version covers the mouth and has holes or prongs for the nares. The total facemask covers the entire face, forming its seal on the forehead, cheeks, and chin. Last, various mouthpiece interfaces can provide positive pressure through an angled tube or straw attached to a flexible arm.

There is no evidence that one interface is superior to others, and often the interface is chosen based on physician or respiratory therapist preference, patient comfort, and symptom-specific needs. For example, a nasal interface is often the preference for nocturnal NPPV in DMD, supplemented by daytime mouthpiece positive pressure ventilation (MPV) (40). For a patient with ALS and bulbar dysfunction, the lips may not form a sufficient seal around a mouthpiece for effective ventilation. Each interface has advantages and disadvantages (**Table 2**). Detailed descriptions of interfaces along with images of their use on patients can be found elsewhere (53, 54).

The use of a nasal interface can result in air leakage out of the mouth and negatively impact patient comfort and sleep architecture (54). For patients who are unable to keep their mouth closed to prevent this, especially during sleep, a chinstrap may be helpful, though different studies have shown variable effectiveness at preventing air leaks with the use of a chin strap (55, 56). Persistent air leak with a nasal interface can be improved with the use of an oronasal, hybrid, or full face mask interface. In some circumstances, patients can alternate interfaces, such as using nasal pillows during the day and an oronasal mask at night. The rotation of interfaces can also reduce the likelihood of skin breakdown, which happens most commonly at the bridge of the nose, the upper lip, and the nasal mucosa (52). Regardless of the selected interface, it is important that sizing is optimized and that straps are sufficiently tightened to minimize air leak while not overly tightened to avoid skin breakdown and maintain comfort.

Ventilator Options and Modes

The use of home noninvasive ventilation has increased dramatically since the efficacy of continuous positive airway pressure (CPAP) to treat obstructive sleep apnea was demonstrated in 1981 (48). CPAP alone is generally not adequate to treat the obstruction and hypoventilation found in NMD, as it fails to address weak muscles, unlike bilevel positive airway pressure or other varieties of ventilation support that can be provided via a mask. Fortunately, various home NPPV device options and modes supporting ventilation are available. The most common devices for NMD provide bilevel support with or without a backup rate. Various modes are available, including pressure support, pressure control, and volume-assured pressure support (VAPS). The most common mode is bilevel with a backup rate (S/T, or spontaneous/timed, mode) (57). In general, devices and modes that offer a backup rate are preferred.

Protocols for initiation of NPPV are often institution dependent. A common approach is to initiate a patient on bilevel support with an expiratory positive airway pressure (EPAP) of

Table 2 Advantages and disadvantages of noninvasive positive pressure ventilation interfaces (adapted from Reference 32)

Interface	Advantages	Disadvantages
Nasal mask	Less aspiration risk Easy secretion clearance Speech and swallowing easier Less claustrophobia Less dead space	Mouth leak Nasal passages have higher resistance Not effective with nasal obstruction Nasal irritation, mouth dryness
Nasal pillow	Eyeglasses can be worn Less skin breakdown Simple setup, easy to fit	Mouth leak Nasal passages have higher resistance Not effective with nasal obstruction Nasal irritation, mouth dryness
Oronasal mask	Better oral leak control More effective in mouth breathers	Claustrophobia Increased aspiration risk Difficult to speak or eat
Hybrid	Less oral leak More effective in mouth breathers Eyeglasses can be worn Less skin breakdown	Increased aspiration risk Difficult to speak or eat
Total face mask	Can be more comfortable Easy to fit Less facial skin breakdown	Risk of rebreathing Eye dryness
Mouthpiece	Less speech interference Low dead space No headgear required May be able to eat	Must maintain mouth seal Usually requires alternative interface during sleep Can cause hypersalivation Potential for orthodontic injury

4–5 cm H_2O and an inspiratory positive airway pressure (IPAP) 4–5 cm H_2O above the EPAP. The backup respiratory rate can be determined by observing the patient's intrinsic respiratory rate and setting a backup rate that is two breaths per minute lower. The pressure support window (the difference between the IPAP and the EPAP) is then adjusted on the basis of the observed tidal volume in order to achieve a target tidal volume of 6–8 mL/kg ideal body weight, which approximates normal breath volume (57, 58). Many devices can estimate obstructive apneas, and EPAP can be titrated to minimize these events. If symptoms or laboratory evidence of hypercapnia develop then the pressure support window can be increased to increase ventilation.

Other modes of ventilation may also be used for chronic respiratory failure. Volume-limited rather than pressure-limited ventilation is possible via noninvasive interface. Historically, patients with ALS were thought to not tolerate volume-limited modes as well as pressure-limited modes (59), though a more recent study suggested that 92% of patients with ALS initiated on volume control continuous mandatory ventilation were tolerating it after 3 months of use (60). VAPS offers a theoretical advantage in patients with progressive NMDs such as ALS, since the mode is designed to maintain a target tidal volume by titrating the pressure support window within a range dictated by the provider. Therefore, as the patient's respiratory muscle strength decreases over time, the device adjusts by offering greater IPAP to maintain a steady tidal volume. A retrospective analysis comparing patients with ALS on bilevel or VAPS modes found that VAPS produced a small but significant increase in tidal volume relative to bilevel, but minute ventilation, respiratory rate, and NPPV daily usage did not differ (61). As of now, no mode of NPPV management has a strong evidence base for important patient-centered outcomes.

MPV is a method of delivering on-demand breaths to a patient utilizing a volume preset ventilator set in the assist-control mode via extension tubing and a mouthpiece interface. Mouthpiece ventilation is considered an excellent option for daytime ventilatory support in NMD where daytime respiratory failure is present and the patient can tolerate the MPV interface (62, 63). Another option is for mask ventilation to be used diurnally, although this can interfere with speech and swallowing (64).

The first home ventilator with a dedicated MPV mode was developed in 2012 (53), although off-label use of home ventilators for this purpose has been available since the 1950s (65). A recent European Neuromuscular Centre International Expert Workshop provided guidelines for the management of MPV in NMD (53, 66). Indications for MPV include, but are not limited to, dyspnea relieved by NPPV, use of mask ventilation >12 h/day, and vital capacity <30% predicted. A notable contraindication in the context of ALS includes the inability to form a lip seal around the mouthpiece. Additionally, caution is advised with hypersalivation (66). Breaths are preferably volume cycled with positive end-expiratory pressure set to zero and are triggered by flow interruption detection, in which the patient interrupts a constant flow from the ventilator by touching the mouthpiece (53). This trigger allows even patients unable to generate an inspiratory effort to use MPV. In addition to providing as-needed ventilatory support while maintaining the ability to speak, MPV allows for breath stacking for cough assistance and lung volume recruitment.

TRACHEOSTOMY HOME VENTILATION

On occasion, TV is necessary for an individual with NMD and chronic neuromuscular respiratory failure. Indications may include patient preference, lack of providers with experience in NPPV, or (most commonly) presence of bulbar involvement, which precludes using either a mask or mouthpiece interface. There is no one test that predicts whether an individual will be able to use a noninvasive interface, and so a trial in the clinic or hospital is the only way to assess this ability. The decision for TV is not an easy one, for the TV will increase the care needs of patients due to the need for tracheostomy changes and suctioning of the airway. Therefore, most expert practitioners in this area strongly advocate for every possible attempt at optimizing NPPV, even for patients requiring 24-h ventilatory support, prior to transitioning to TV (67). In a survey of patients with ALS and their caregivers, 81% of patients who chose tracheostomy with mechanical ventilation said that they would do so again and that they would advise other patients to do the same. However, 30% of caregivers rated the patient's overall quality of life worse than their patient did, and only 50% would opt for ventilation themselves. In contrast, among caregivers of those on NPPV, 94% would opt for NPPV themselves (68).

There is no optimal mode of TV and any volume- or pressure-regulated mode can be used and titrated to the patient's needs. TV is considered only for patients who would like aggressive respiratory support and cannot continue with NPPV, for whom the alternative would likely be death.

AIRWAY CLEARANCE

The normal cough strength and coordination, along with the mucociliary clearance of the airways, are impaired in patients with NMD (69). In order to have an effective cough (70), a patient needs intact inspiratory strength, expiratory strength, and glottic closure coordination. As described earlier, a deficiency of any of these leads to a weakened cough (26). Ineffective cough and progressive bulbar disease are the main causes of mortality for patients with NMD (69).

Several adjunctive therapies can improve cough efficacy. Cough augmentation is generally achieved through increasing inspiratory volume, accomplished with a variety of techniques: use

of a lung volume recruitment bag, a noninvasive device, or glossopharyngeal breathing (69, 71). These all assist with taking single or multiple sequential breaths to increase vital capacity and peak cough flow. A mechanical insufflation device is specifically built for this purpose. In addition, the expiratory limb of a cough can also be assisted with a negative pressure device, often a combined maximal insufflation-exsufflation (MI-E) device (71). One important limitation to the MI-E is in patients with bulbar dysfunction, who may experience dynamic collapse of the upper airways during the exsufflation cycle, thus limiting the usefulness of the device (72).

Other adjuncts target mucus clearance through mechanical and chemical disruption of the mucus itself. The use of manual techniques such as abdominal thrusts, flutter devices, external oscillatory and compression devices (often vests), and pharmaceuticals aimed at thinning mucus might all be employed (69).

UNMET NEEDS

Currently, there are several unmet needs for the care of patients with chronic respiratory failure due to NMD. In addition to an insufficient number of skilled practitioners (clinicians, respiratory therapists, and nurses, among others), there is significant variability in the availability and financing of devices and care needed (66). Patients with NMD uniquely require significant care, much of which is provided by family members. Care assistance is expensive, and analyses of cost effectiveness are lacking (73).

CONCLUSIONS

Chronic respiratory failure is an important cause of morbidity and mortality in patients with chronic NMD. The impairment of inspiratory, expiratory, and bulbar muscles leads to long-term respiratory failure, with hallmark features of hypercapnia, dyspnea, and poor mucus and secretion management. The management of chronic respiratory failure in this context requires multimodal approaches to these issues, with noninvasive positive pressure as the foundation for ventilatory support.

DISCLOSURE STATEMENT

J.O.B. has served on the Medical Advisory Board for Ventec Life Systems, a home ventilator manufacturer.

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