Journal of Pulmonary & Respiratory Medicine

Comer, J Pulm Respir Med 2014, 5:1
DOI: 10.4172/2161-105X.1000234

Review Article Open Access

An Update on Domiciliary Non-Invasive Ventilation

Comer DM*

Academic Department of Sleep & Ventilation, Directorate of Respiratory Medicine & Physiology, Birmingham Heartlands Hospital, UK

Abstract

Chronic respiratory failure (CRF) should be considered a syndrome defined by arterial blood gas abnormalities. Domiciliary non-invasive ventilation (NIV) is an increasingly used intervention to curtail the detrimental effects of CRF in individuals with a broad range of cardiorespiratory disorders. Progress in the provision, monitoring and improvement in patient centred outcomes with NIV has simply been staggering over the last decade. Despite this, questions still remain in terms of the ideal mode of delivery, the most efficient techniques to monitor its effectiveness, and the timing of NIV initiation for several of the more common diseases which can require NIV support.

There are a number of mechanisms accounting for the reduced ventilation that is apparent during sleep, even in normal subjects. This represents a particularly unique physiological state whereby changes in upper airway resistance control of respiration and changes in lung mechanics have an impact on the adequacy of tidal volume breathing. Abnormal respiratory events tend to occur specifically during sleep, and so NIV which is appropriate for the awake patient may not be suitable when asleep at night. These factors need to be kept in mind both for the optimal timing of NIV, and the manner in which it should be monitored.

This article aims to provide an overview of current concepts in the appropriate mode of delivery of NIV, discusses the patient groups who benefit from its application, highlights challenges with interface design, and aspects that need to be kept in mind in terms of the optimal monitoring of NIV.

Keywords: Chronic respiratory failure (CRF); Non-invasive ventilation; Non-rapid eye-movement; Congestive heart failure

Introduction

The purpose of domiciliary non-invasive ventilation (NIV), in its essence, is to mitigate nocturnal hypoventilation, control any associated symptoms, and to strive to achieve adequate and consistent nocturnal oxygen saturation. During non-rapid eye-movement (NREM) sleep, during which time the impetus to breath becomes more dependent on the metabolic control of breathing, there tends to be a heightened vulnerability to apnoeas and hypopnoeas. Furthermore, a decrement in minute ventilation can develop, not only as a result of airway compromise, but also as a consequence of the overall increase in the work of breathing and changes in mechanics of breathing itself. During rapid eye-movement sleep (REM), on the other hand, a reduction in accessory muscle contraction occurs in conjunction with an erratic respiratory drive. These changes can ultimately result in the development of apnoeas, hypopnoeas, alveolar hypoventilation, and ventilation perfusion mismatching (Figure 1) [1].

Progress in the provision, ease of application, and improved monitoring of the efficacy of NIV, which itself aims to address the aforementioned abnormalities, has simply been staggering over the last decade. This progress can be attributed not only to the increasing number of indications in which the effectiveness of NIV has been established, but in addition, to the availability of high-performance ventilators and to the development of a strong technical support infrastructure.

As well as an improvement in nocturnal gas exchange, there is in additional rationale for NIV to provide a sustained period of rest for the muscles of respiration, particularly for those subjects who have a heightened work of breathing when asleep. This is based on ameliorating the increased work of breathing while asleep, and so improving muscle strength, contractility, and overall ventilatory mechanics, and, as a consequence, reducing the decline in muscle strength and so improving overall functional ability in patients who have primarily neuromuscular weakness [2]. Interestingly, the use of domiciliary NIV during the daytime, and so not necessarily sleep related, has been shown to have a similar efficacy as compared to nocturnal NIV on daytime physiological measurements in patients with chronic hypoventilation [3-8].

Patient Groups

A broader range of subjects with a variety of cardio-respiratory disorders are now supported with NIV than was the case historically. Physiological parameters, level of patient motivation, other co-morbidities, and the prognosis of the disease itself all need to be taken into consideration when the decision to proceed with long term domiciliary NIV for any given individual. All of these aforementioned elements are closely related to each other, and it can be challenging to disentangle these elements completely in an effort to meaningfully conclude whether, in isolation, any aspect should influence the decision as to the appropriateness or otherwise to commence domiciliary NIV. With increasing age of patients who are now treated for respiratory failure, these decisions can at times become particularly complex.

The neuromuscular diseases which can often lead to respiratory insufficiency, such as duchenne muscular dystrophy (DMD) and amyotrophic lateral sclerosis (ALS), along with chronic obstructive pulmonary disease (COPD), are well known to predispose individuals to sleep hypoventilation and ventilatory insufficiency, and the rationale for the application of NIV, particularly in the acute setting, is simply beyond reproof. In more recent times, and perhaps somewhat less intuitively, is the rationale for NIV to improve awake hypoventilation by providing ventilatory support for sleep-related disordered breathing

***Corresponding author**: Comer DM MRCP, PhD, Acting Consultant, Sleep & Ventilation Medicine, Academic Department of Sleep & Ventilation, Directorate of Respiratory Medicine & Physiology, Birmingham Heartlands Hospital, Bordesley Green East, Birmingham B9 5SS, UK, Tel: ++44-0-2892640654; E-mail: dcomer@doctors.org.uk

Received January 12, 2015; **Accepted** January 30, 2015; **Published** February 03, 2015

Citation: Comer DM (2015) An Update on Domiciliary Non-Invasive Ventilation. J Pulm Respir Med 5: 234. doi:10.4172/2161-105X.1000234

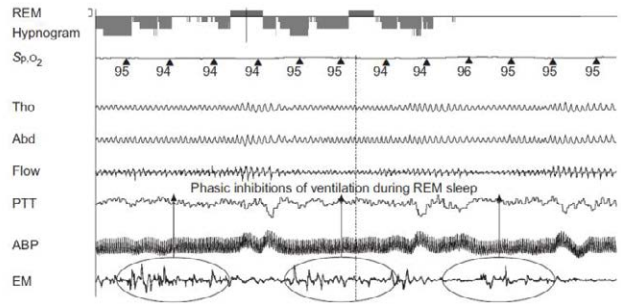

Figure 1: Irregular, rapid and shallow breathing during phasic rapid eye movement (REM) sleep. Decrease in ventilation during phasic REM sleep episodes: a sustained reduction in flow and thoracic and abdominal movements occurs concurrently with rapid eye movements. This occurs to various degrees in normal subjects and is aggravated or revealing in patients with respiratory failure. SpO_2: Arterial oxygen saturation measured by pulse oximetry; Tho: Thoracic movements; Abd: Abdominal movements; Flow: Flow signal; PTT: Pulse transit time; ABP: Arterial blood pressure; EM: Eye movements. Reproduced with permission of the European Respiratory Society©.Eur Respir Mon November 2008 41:350-366.

by interrupting the vicious cycle of chronic hypercapnia-associated blunting of chemosensitivity and ventilatory drive, along with a reduction in sleep associated hypercapnia, acidosis and hypoxia-associated impairment in respiratory muscle and peripheral nerve function [5,9].

The periodic breathing characteristic of congestive heart failure (CHF) commonly referred to as Cheyne-Stokes breathing, has a cyclical crescendo-decrescendo pattern of respiratory effort and airflow, with or without outright central sleep apnoea at the nadir of the breathing cycle, and in the absence of upper airway obstruction. Central sleep apnoea, in this context, is believed to be as a result of delayed chemoreceptor responses as a consequence of an increased circulatory time. Individuals with CHF often have a baseline hypocapnia both during wakefulness and sleep [10,11]. The apnoea during sleep as a consequence of the low PaCO2, with the subsequent delay in the sensing of the resulting increased PaCO2, leads to the typical crescendo-descrendo pattern typical of hyperpnoea with arousal, and subsequent hypopnoea and apnoea. All of these effects appear to be most pronounced at times of hospitalisation for decompensated heart failure [12].

The spectrum of neuromuscular disorders which potentially benefit from domiciliary NIV is broad. Indications include isolated diaphragmatic paralysis, the muscular dystrophies, ALS, neuromuscular junction disease (such as myasthenia gravis), demyelinating diseases, and the various spinal cord disorders. The timing of commencement of NIV for DMD patients is critical. For those individuals who demonstrate evidence of sleep disordered breathing, NIV is clearly of benefit [13]. On the other hand, inappropriately applied NIV can potentially lead to patient-ventilator dyssynchrony and so lead to impairment, rather than enhancement, of ventilatory function. One study demonstrated a decreased long-term survival in DMD subjects with respiratory insufficiency using domiciliary NIV when compared to controls in the absence of awake hypercapnia and sleep disordered breathing [14].

ALS is a form of neuromuscular disease which is characterised by a progressive muscular atrophy through the body. Hypoventilation, as a direct consequence of this muscle weakness, is the most common cause of death in advanced ALS cases [15,16]. Bulbar symptoms and age of 65 years or greater at the time of diagnosis are poor prognostic factors [17]. Treatment interventions are palliative. Published guidelines from the American Academy of Neurology indicates, for individuals with ALS, that domiciliary NIV has a beneficial role in extending survival, mitigating the decline in forced vital capacity, relieving dyspnoea and improving quality of life [18]. Unfortunately many ALS patients with bulbar dysfunction are not able to tolerate NIV [19] although for this group NIV still can confer improved symptoms and quality of life, particularly for those subjects who are hypercapnic at initiation of NIV [20]. Interestingly, these guidelines do not specifically state that NIV should be given nocturnally, although this is the most conventional time, even in the absence of evidence of sleep-related breathing disorders in an individual subject.

In addition to the routine monitoring of respiratory symptoms, the ALS guidelines indicate that nocturnal oximetry and maximal inspiratory pressure are the most sensitive monitoring measures. Reliance on clinical symptoms alone, in the context of ALS, is not the most sensitive means of recognising respiratory muscle weakness, and the degree of respiratory impairment can be easily underestimated. In one particular study, of 45 subjects with a forced vital capacity less than 50%, the attending neurologist considered respiratory muscle to be normal in 29, and only mildly impaired in 15 subjects [21].

Initially the use of NIV, as a form of supportive therapy, was reserved almost exclusively for subjects with acute exacerbations of chronic obstructive pulmonary disease (AECOPD), and avoidance of tracheal intubation was achieved for many of these particular subjects [22]. A series of randomised controlled trials have demonstrated that the addition of NIV to standard therapy reduces levels of dyspnoea in addition to a reduction in length of hospital stay [23-26]. However, in the domiciliary setting, and in the more long term setting, the role of NIV is more controversial. This is reflected in the fact that the use of domiciliary NIV for subjects with COPD varies across Europe, with the highest prevalence in Italy and the lowest prevalence in the Scandinavian countries [27].

Initial trials published in the 1990s did not report benefits of domiciliary NIV for people with COPD. However, these trials have been criticised by a lack of stringent criteria in terms of ensuring only those subjects with an episode of hypercapnic respiratory failure were enrolled, and it has been suggested that insufficient pressures were often used, resulting in a poor patient selection and poor adherence to domiciliary NIV [28-32]. In contrast, a smaller number of trials have demonstrated more favourable outcomes, indicating that domiciliary NIV for COPD results in an improvement in gas exchange, sleep quality and exercise capacity, when domiciliary NIV was added to LTOT [33,34]. Interestingly, recent work indicated that although combining domiciliary NIV with a formal exercise training programme augmented the beneficial effects of each modality in isolation when gas exchange and selected biomarkers were considered, the BODE index, dyspnoea levels (determined by the modified Medical Research Council dyspnoea scale) and quality of life (determined by the Chronic Respiratory Disease Questionnaire) were similar between the study groups [35].

The differences in the methodology of these latter studies would at least suggest that higher NIV pressures should be adopted for supporting respiratory insufficiency in the context of COPD, a notion which is supported by recent randomised cross-over trials of NIV in COPD [36,37]. The most recent RCT trial published from a German group investigating the role of domiciliary NIV in COPD using high inspiratory pressures recently reported that NIV improved survival when settings were targeted to substantially reduce hypercapnia by 20% of below 6.5 kPa [38]. A similar trial is currently underway in the UK (NCT Identifier: NCT00990132).

NIV Strategies

The selection of the most appropriate mode of NIV must be based upon patient characteristics and the ventilatory assist should be tailored to individual patient characteristics keeping in mind the underlying cardiorespiratory disorder. For bilevel ventilation, the expiratory positive airway pressure (EPAP) is set so as to overcome upper airway resistance and to maintain airway patency, while the inspiratory positive airway pressure (IPAP) is set at a level above the EPAP to generate a sufficient pressure gradient to overcome the inherent resistance of the entire respiratory circuit (including the ventilatory apparatus) and also generate a target ventilation. A respiratory rate is may be spontaneous alone or with a mandated "back up" rate, or a combination of both.

In more general terms, pressure and volume targeted ventilatory targets are used in chronic respiratory failure, both of which are efficacious when short term gas exchange end points are considered [39-41]. However, pressure support ventilation has less gastrointestinal side effects and demonstrates enhanced comfort in comparison to volume targeted devices [42-45]. Other more adaptive modes of ventilation, such as proportional assist ventilation which delivers a pressure proportional to inspiratory effort [46]. Adaptive servo ventilation, and average volume assured pressure support are also available.

The advantage of volume targeted ventilation is that the inspiratory volume, and so the tidal volume achieved, remains relatively stable. However, in order to achieve this, there can be significant variation in IPAP levels which can prove challenging for individual patients, and any leakages which are a common problem with NIV are not satisfactorily compensated for. Furthermore, the delivery of the relatively fixed tidal volume does not permit account to be taken of the patients' varying requirements. On the other hand, the tidal volume delivered by pressure support ventilation may vary in the context of a heightened airway resistance. Any leaks during the delivery of pressure support ventilation are generally well compensated for by variations in the delivered pressure. Each breathing cycle is terminated by a flow criterion rather than by volume or time, and the patient retains control of the cycle length in addition to the depth and overall flow profile. Pressure support ventilators are cheaper. With all of these aspects in mind, it is perhaps not particularly surprising that pressure support ventilation dominates the domiciliary ventilation arena. It is however important for health care providers to be cognisant of the advantages and disadvantages of the two modalities, and make a balanced approach as to the most appropriate modality for any given patient.

Classically, NIV ventilators have two types of triggers. The pressure based trigger, mainly present in older ventilators, is based on the detection of an inspiratory flow within the circuit. This has generally been superseded by a flow based trigger, which are more universal, and depends on detection of inspiratory flow in the context of continuous flow within the circuit itself. The duration between respiratory effort and delivery of the delivered tidal breath, along with how appropriately trigger sensitivity is adjusted for inspiratory effort, are elements which can contribute to ventilator asynchrony, and in theory, potentially disrupt sleep quality.

The requirements of patients with different conditions may vary, and so studying groups of patients which comprise a broad variety of disease states may mask important differences which are of only of relevance to particular patient groups. Studies have shown equivalence [42,47-49] benefit of pressure [50] or benefit of volume [51], whereas other more long term studies have been divergent in their findings, or have used a heterogeneous group of patients [42,47,52].

It should be kept in mind that the ventilator settings were not consistently equivalent throughout these studies, and so their findings are not truly assessing any difference in the mode of delivery alone in terms of the efficacy of ventilation. However, one particular study compared these two particular modes in patients with chest wall deformities using identical volumes of delivered ventilation concluded that pressure and volume ventilation were equivalent in terms improvement in respiratory physiology and the consequent changes in health status [49]. However, it could be speculated that volume targeted ventilation would be more appropriate in other diseases, such as severe neuromuscular disease, which may facilitate patients with impaired cough to breath stack. Levels of leakage have been demonstrated to be higher for pressure when compared to volume controlled ventilation for any given volume of ventilation achieved, although this does not translate into a higher number of arousals, deterioration in quality of life, or reduction in daytime function [49].

More recently, a hybrid type of ventilator has emerged. These are designed in order to overcome the disadvantages of volume and pressure ventilators. The most thoroughly studied mode is the average volume assured pressure support (AVAPS). Initial trails were favourable, indicating that AVAPS mode delivered a more optimal control of nocturnal ventilation when compared to pressure control ventilation for subjects with obesity hypoventilation [53]. However, this was not reflected in improvements in health related quality of life and sleep quality, determined by the Severe Respiratory Insufficiency Questionnaire and polysomnography respectively [54].

A series of subsequent studies comparing AVAPS and pressure support ventilation for subjects with obesity hypoventilation and COPD indicated comparable outcomes for the two modes of ventilation,[37,55,56] although a smaller number of studies reported significantly improved nocturnal hypoventilation with AVAPS [53,57]. The largest randomised controlled trial to date demonstrated equivalence of AVAPS and pressure support ventilation for subjects with obesity hypoventilation when gas exchange, quality of life, and weight loss were considered [58].

Interestingly, for patients with COPD, pressure controlled ventilation, whereby the majority of the delivered breaths occur without triggering and with higher inspiratory pressures, has been demonstrated to be superior to assisted ventilation, requiring a trigger by the patient, in terms of controlling nocturnal hypoventilation and improving health related quality of life [36,59]. However, in terms of the mode of ventilation itself, and considering the evidence overall at this time, neither AVAPS or pressure support ventilation has clearly emerged as consistently more beneficial for domiciliary NIV, and firm recommendations cannot be established.

An appropriate pressurisation rate is vital in order to minimise inspiratory effort and to optimise synchronisation. During this initial phase inspiratory flow should ideally be adequate to most closely match inspiratory demand [60]. The level of ventilatory support, the duration of time required to reach the target pressure, and the overall compliance and resistance of the respiratory system, along with patient inspiratory effort have an impact on his rate. A swift rise time unloads respiratory muscles, although this has the potential to increase the sensation of dyspnoea, [61] can induce double triggering and so increase the chances of leaks around the interface [62]. A slower rise time, on the other hand, increases the inspiratory work required of the patient (Figure 2) [60].

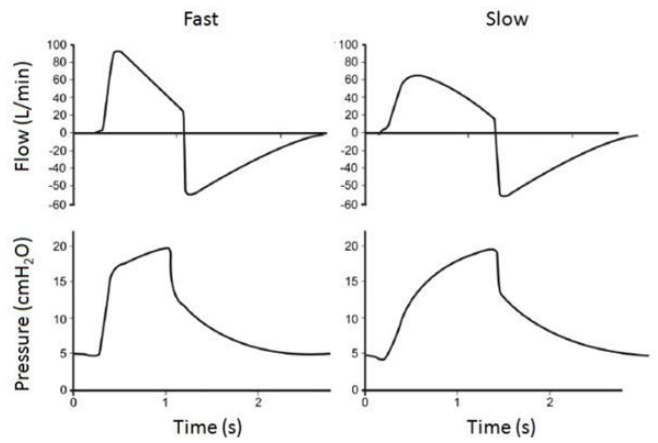

Figure 2: Flow and pressure waveforms for 2 rise times (pressurization rates) at a pressure support of 20 cm H_2O. Note the effect of rise time on flow at the initiation of the inspiratory phase.

NIV Interface

There is no perfect interface for NIV. The full face mask (FFM) is generally regarded as the most appropriate first-line strategy for the management of acute hypercapnic respiratory failure with NIV. In this context, the FFM leads to more favourable patient outcomes, such as a lower probability of progression to tracheal intubation, improved tolerance, and more satisfactory gas exchange when compared to the various nasal masks (NM) [63]. However, for chronic stable respiratory failure, the most commonly chosen interface used is generally regarded as the NM, followed by nasal pillows, primarily on the basis of patient comfort [64,65]. For both scenarios the ideal interface minimises air leaks, maximises patient comfort, and ensures satisfactory patient-ventilator synchrony. Considerable technological progress over the last decade has broadened the number and types of interface to achieve these aims [64].

A variety of interfaces are now available for optimising the efficacy of NIV. These include nasal pillows, nasal masks, oral-nasal masks, mouthpieces and helmets [64]. Air leak through the mouth, a clear challenge with the nasal interfaces, can be mitigated with the use of a chin strap, or failing that, a full face mask. There is evidence to support the notion that heated humidification is beneficial in terms of reducing nasal and/or oral dryness [66].

It is widely accepted that the dead space rendered by virtue of the internal volume of the interface itself is a considerable problem, and this varies considerably between the various interfaces available. Dead space increases with masks which have a larger internal volume [67], whereas in contrast, is decreased in masks which have a built-in exhalation port, ideally located over the nasal bridge [65,68]. These factors have historically rendered nasal interfaces to be considered the most appropriate option for stable chronic patients requiring domiciliary NIV [69].

Monitoring NIV

Appropriate strategies to ensure adequacy of ventilation and respiratory support is necessary. During sleep itself, and respiratory events can be attributed to either sleep related physiological events, or alternatively, due to events as a consequence of the inherent use of NIV in and of itself [70]. The latter can be precipitated by interface leaks, instability of the upper airway, and any residual obstructive events, with or without glottis closure, or patient-ventilator asynchrony (Figure 3).

With this in mind, simply increasing the delivered tidal volume or increasing the inspiratory pressure during NIV does not necessarily result in a more effective tidal volume. Nocturnal monitoring of NIV in the context of respiratory insufficiency is much more challenging when compared to that for CPAP and sleep apnoea. Sleep is associated with profound ventilatory changes, which can be heightened by interactions with the ventilator. Finally, the physicians caring for these subjects adopt varying approaches to establish the effectiveness of NIV which vary from isolated blood gas measurements to full polysomonography [71].

Patient ventilatory asynchrony can be manifested as problems with triggering of the ventilator. This can include ineffective triggering, double triggering, auto-triggering (usually caused by leaks causing the EPAP level to fall below the trigger threshold), and/or with timing out-of-phase. Adjusting trigger and cycling sensitivity, inspiratory time limits and rise times, and time in inspiratory pressure assist as a percentage of the respiratory cycle all serve to optimise efficacy and minimise patient-ventilator asynchrony [72,73]. Some more recent ventilators propose automated complex trigger algorithms by which the actual flow-time waveform is used as a means to identify the appropriate time to trigger the ventilator. Other challenges of NIV, albeit from studies of NIV use in the acute setting, include eye irritation, nasal and oral dryness, aerophagia and gastric distention, vomiting, pneumothorax and pneumonia, among others [74,75].

Patient ventilatory asynchrony is frequent in current standard practice. A large systematic investigation of the physiologic efficacy of NIV for individuals with ALS demonstrated that 17% of the nocturnal polysomnographic recording time was spent in dyssynchrony with the ventilator [76]. This is also a problem in COPD subjects requiring domiciliary NIV [77,78]. Interestingly, when detailed physiological assessments including polysomnography, surface diaphragm electromyography and transcutaneous carbon dioxide measurements are used in order to optimise patient ventilatory synchrony, this leads to a significant reduction in patient reported symptoms of hypoventilation [79]. This is most intriguing, and at least suggests that optimal set up of the ventilator could serve to minimise adverse clinical symptoms and so potentially reduce inadequate adherence.

Options for monitoring purposes can include oxygen desaturation or an elevation in transcutaneous carbon dioxide levels. More modern home ventilators are designed to be capable of detecting and recording a diverse range of parameters over several months and so providing relevant information to the clinician. These include the establishing the amount of leak, if any, degree of compliance, overall use in hours per night, tidal volumes achieved, and for some ventilators, an estimate of persistent respiratory compromise in the form of an apnoea hypopnoea index. In selected patients a more complete evaluation with polysomnographic recordings is necessary who have persistent symptoms despite satisfactory compliance and perceived optimal ventilator settings (Figure 4).

Conclusion

Domiciliary NIV improves gas exchange, oxyhaemoglobin saturation and symptoms of hypoventilation in a broad range of cardiorespiratory disorders. Despite this, and perhaps a little surprisingly, there are, relatively speaking, few high quality data on the impact of domiciliary NIV in terms of physiological responses. Questions remain in terms of benefits of long term survival, quality of life, and patient centred outcomes in a number of disease states.

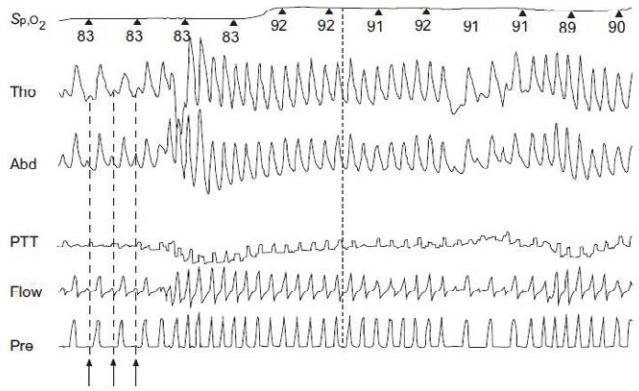

Figure 3: 5-min epoch of polysomnography with patient–ventilator asynchrony. Ineffective efforts (arrows) and arterial oxygen saturation oscillations related to patient–ventilator asynchrony. SpO2: arterial oxygen saturation measured by pulse oximetry; Tho: thoracic movements; Abd: abdominal movements; PTT: pulse transit time; Flow: flow signal; Pre: pressure. Reproduced with permission of the European Respiratory Society©. Eur Respir Mon November 2008 41:350-366.

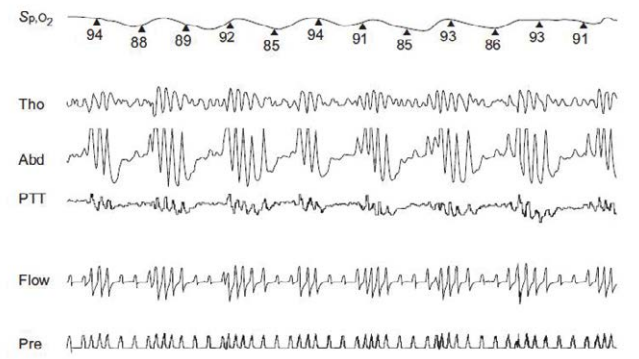

Figure 4: 5-min epoch of polysomnography with central events induced by noninvasive ventilation. Arterial oxygen saturation measured by pulse oximetry (SpO2) oscillations related to central events occurring in stage-1 sleep. Note the decrease in respiratory effort during events, identified by pulse transit time (PTT). Tho: thoracic movements; Abd: Abdominal movements; Flow: Flow signal measured using a pneumotachograph; Pre: Pressure. Reproduced with permission of the European Respiratory Society©. Eur Respir Mon November 2008 41:350-366.

Furthermore, there is a need for more robust data with regard to which disorders primarily benefit from NIV applied during sleep as opposed to awake, particularly for those subjects who do not have a specific sleep-related breathing disorder. This is of relevance to those individuals who have difficulty tolerating the NIV, demonstrate high levels of patient ventilator asynchrony, or who may more efficiently use NIV when awake in an effort to improve respiratory muscle mechanics.

Despite these shortcomings in our knowledge, long term domiciliary NIV does have the capacity to reduce hypercapnia, prolong survival, and to reduce hospital admission frequency. This form of supportive therapy is most efficacious when offered to the right patient, at the right time, and in the right way.

References

1. Smith PE, Edwards RH, Calverley PM (1989) Ventilation and breathing pattern during sleep in Duchenne muscular dystrophy. Chest 96: 1346-1351.
2. Renston JP, DiMarco AF, Supinski GS (1994) Respiratory muscle rest using nasal BiPAP ventilation in patients with stable severe COPD. Chest 105: 1053-1060.
3. Heckmatt JZ, Loh L, Dubowitz V (1990) Night-time nasal ventilation in neuromuscular disease. Lancet 335: 579-582.
4. Gutiérrez M, Beroíza T, Contreras G, Díaz O, Cruz E, et al. (1988) Weekly cuirass ventilation improves blood gases and inspiratory muscle strength in patients with chronic air-flow limitation and hypercarbia. Am Rev Respir Dis 138: 617-623.
5. Barbé F, Quera-Salva MA, de Lattre J, Gajdos P, Agustí AG (1996) Long-term effects of nasal intermittent positive-pressure ventilation on pulmonary function and sleep architecture in patients with neuromuscular diseases. Chest 110: 1179-1183.
6. Scano G, Gigliotti F, Duranti R, Spinelli A, Gorini M, et al. (1990) Changes in ventilatory muscle function with negative pressure ventilation in patients with severe COPD. Chest 97: 322-327.
7. Shneerson JM (1996) The changing role of mechanical ventilation in COPD. Eur Respir J 9: 393-398.
8. Priou P, Hamel JF, Person C, Meslier N, Racineux JL, et al. (2010) Long-term outcome of noninvasive positive pressure ventilation for obesity hypoventilation syndrome. Chest 138: 84-90.
9. Mayer P, Dematteis M, Pépin JL, Wuyam B, Veale D, et al. (1999) Peripheral neuropathy in sleep apnea. A tissue marker of the severity of nocturnal desaturation. Am J Respir Crit Care Med 159: 213-219.
10. Hanly P, Zuberi N, Gray R (1993) Pathogenesis of Cheyne-Stokes respiration in patients with congestive heart failure. Relationship to arterial PCO2. Chest 104: 1079-1084.
11. Lorenzi C, Berthommier F, Apoux F, Bacri N (1999) Effects of envelope expansion on speech recognition. Hear Res 136: 131-138.
12. Padeletti M, Green P, Mooney AM, Basner RC, Mancini DM (2009) Sleep disordered breathing in patients with acutely decompensated heart failure. Sleep Med 10: 353-360.
13. Segall D (1988) Noninvasive nasal mask-assisted ventilation in respiratory failure of Duchenne muscular dystrophy. Chest 93: 1298-1300.
14. Raphael JC, Chevret S, Chastang C, Bouvet F (1994) Randomised trial of preventive nasal ventilation in Duchenne muscular dystrophy. French Multicentre Cooperative Group on Home Mechanical Ventilation Assistance in Duchenne de Boulogne Muscular Dystrophy. Lancet 343: 1600-1604.
15. Tandan R, Bradley WG (1985) Amyotrophic lateral sclerosis: Part 1. Clinical features, pathology, and ethical issues in management. Ann Neurol 18: 271-280.
16. Rowland LP, Shneider NA (2001) Amyotrophic lateral sclerosis. N Engl J Med 344: 1688-1700.
17. del Aguila MA, Longstreth WT Jr, McGuire V, Koepsell TD, van Belle G (2003) Prognosis in amyotrophic lateral sclerosis: a population-based study. Neurology 60: 813-819.
18. Miller RG, Jackson CE, Kasarskis EJ, England JD, Forshew D, et al. (2009) Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: drug, nutritional, and respiratory therapies (an evidence-based review): report of the Quality Standards Subcommittee of the American Academy of Neurology. Neurology 73: 1218-1226.
19. Siirala W, Aantaa R, Olkkola KT, Saaresranta T, Vuori A (2013) Is the effect of non-invasive ventilation on survival in amyotrophic lateral sclerosis age-dependent? BMC Palliat Care 12: 23.
20. Bourke SC, O'Neill CL, Williams TL, Peel ET, Gibson GJ, et al. (2012) The changing landscape of non-invasive ventilation in amyotrophic lateral sclerosis. J Neurol Neurosurg Psychiatry 83: 368-369.
21. Parreira VF, Jounieaux V, Aubert G, Dury M, Delguste PE, et al. (1996) Nasal two-level positive-pressure ventilation in normal subjects. Effects of the glottis and ventilation. Am J Respir Crit Care Med 153: 1616-1623.
22. Meduri GU, Conoscenti CC, Menashe P, Nair S (1989) Noninvasive face mask ventilation in patients with acute respiratory failure. Chest 95: 865-870.
23. Plant PK, Owen JL, Elliott MW (2000) Early use of non-invasive ventilation for acute exacerbations of chronic obstructive pulmonary disease on general respiratory wards: a multicentre randomised controlled trial. Lancet 355: 1931-1935.
24. Brochard L, Mancebo J, Wysocki M, Lofaso F, Conti G, et al. (1995) Noninvasive ventilation for acute exacerbations of chronic obstructive pulmonary disease. N Engl J Med 333: 817-822.

25. Conti G, Antonelli M, Navalesi P, Rocco M, Bufi M, et al. (2002) Noninvasive vs. conventional mechanical ventilation in patients with chronic obstructive pulmonary disease after failure of medical treatment in the ward: a randomized trial. Intensive Care Med 28: 1701-1707.

26. Bott J, Carroll MP, Conway JH, Keilty SE, Ward EM, et al. (1993) Randomised controlled trial of nasal ventilation in acute ventilatory failure due to chronic obstructive airways disease. Lancet 341: 1555-1557.

27. Lloyd-Owen SJ, Donaldson GC, Ambrosino N, Escarabill J, Farre R, et al. (2005) Patterns of home mechanical ventilation use in Europe: results from the Eurovent survey. Eur Respir J 25: 1025-1031.

28. Casanova C, Celli BR, Tost L, Soriano E, Abreu J, et al. (2000) Long-term controlled trial of nocturnal nasal positive pressure ventilation in patients with severe COPD. Chest 118: 1582-1590.

29. Gay PC, Hubmayr RD, Stroetz RW (1996) Efficacy of nocturnal nasal ventilation in stable, severe chronic obstructive pulmonary disease during a 3-month controlled trial. Mayo Clin Proc 71: 533-542.

30. Strumpf DA, Millman RP, Carlisle CC, Grattan LM, Ryan SM, et al. (1991) Nocturnal positive-pressure ventilation via nasal mask in patients with severe chronic obstructive pulmonary disease. Am Rev Respir Dis 144: 1234-1239.

31. Lin CC (1996) Comparison between nocturnal nasal positive pressure ventilation combined with oxygen therapy and oxygen monotherapy in patients with severe COPD. Am J Respir Crit Care Med 154: 353-358.

32. McEvoy RD, Pierce RJ, Hillman D, Esterman A, Ellis EE, et al. (2009) Nocturnal non-invasive nasal ventilation in stable hypercapnic COPD: a randomised controlled trial. Thorax 64: 561-566.

33. Clini E, Sturani C, Rossi A, Viaggi S, Corrado A, et al. (2002) The Italian multicentre study on noninvasive ventilation in chronic obstructive pulmonary disease patients. Eur Respir J 20: 529-538.

34. Meecham Jones DJ, Paul EA, Jones PW, Wedzicha JA (1995) Nasal pressure support ventilation plus oxygen compared with oxygen therapy alone in hypercapnic COPD. Am J Respir Crit Care Med 152: 538-544.

35. Márquez-Martín E, Ruiz FO, Ramos PC, López-Campos JL, Azcona BV, et al. (2014) Randomized trial of non-invasive ventilation combined with exercise training in patients with chronic hypercapnic failure due to chronic obstructive pulmonary disease. Respir Med 108: 1741-1751.

36. Dreher M, Storre JH, Schmoor C, Windisch W (2010) High-intensity versus low-intensity non-invasive ventilation in patients with stable hypercapnic COPD: a randomised crossover trial. Thorax 65: 303-308.

37. Murphy PB, Brignall K, Moxham J, Polkey MI, Davidson AC, et al. (2012) High pressure versus high intensity noninvasive ventilation in stable hypercapnic chronic obstructive pulmonary disease: a randomized crossover trial. Int J Chron Obstruct Pulmon Dis 7: 811-818.

38. Köhnlein T, Windisch W, Wegscheider K, Welte T (2014) Non-invasive positive pressure ventilation for severe COPD--Authors' reply. Lancet Respir Med 2: e19.

39. Restrick LJ, Fox NC, Braid G, Ward EM, Paul EA, et al. (1993) Comparison of nasal pressure support ventilation with nasal intermittent positive pressure ventilation in patients with nocturnal hypoventilation. Eur Respir J 6: 364-370.

40. Chadda K, Clair B, Orlikowski D, Macadoux G, Raphael JC, et al. (2004) Pressure support versus assisted controlled noninvasive ventilation in neuromuscular disease. Neurocrit Care 1: 429-434.

41. Muñoz X, Crespo A, Marti S, Torres F, Ferrer J, et al. (2006) Comparative study of two different modes of noninvasive home mechanical ventilation in chronic respiratory failure. Respir Med 100: 673-681.

42. Windisch W, Storre JH, Sorichter S, Virchow JC Jr (2005) Comparison of volume- and pressure-limited NPPV at night: a prospective randomized cross-over trial. Respir Med 99: 52-59.

43. Vitacca M, Rubini F, Foglio K, Scalvini S, Nava S, et al. (1993) Non-invasive modalities of positive pressure ventilation improve the outcome of acute exacerbations in COLD patients. Intensive Care Med 19: 450-455.

44. Hess DR (2004) The evidence for noninvasive positive-pressure ventilation in the care of patients in acute respiratory failure: a systematic review of the literature. Respir Care 49: 810-829.

45. Pinto A, Almeida JP, Pinto S, Pereira J, Oliveira AG, et al. (2010) Home telemonitoring of non-invasive ventilation decreases healthcare utilisation in a prospective controlled trial of patients with amyotrophic lateral sclerosis. J Neurol Neurosurg Psychiatry 81: 1238-1242.

46. Hess DR (2006) Noninvasive ventilation in neuromuscular disease: equipment and application. Respir Care 51: 896-911.

47. Elliott MW, Aquilina R, Green M, Moxham J, Simonds AK (1994) A comparison of different modes of noninvasive ventilatory support: effects on ventilation and inspiratory muscle effort. Anaesthesia 49: 279-283.

48. Meecham Jones DJ, Wedzicha JA (1993) Comparison of pressure and volume preset nasal ventilator systems in stable chronic respiratory failure. Eur Respir J 6: 1060-1064.

49. Tuggey JM, Elliott MW (2005) Randomised crossover study of pressure and volume non-invasive ventilation in chest wall deformity. Thorax 60: 859-864.

50. Lien TC, Wang JH, Huang SH, Chen SD (2000) Comparison of bilevel positive airway pressure and volume ventilation via nasal or facial masks in patients with severe, stable COPD. Zhonghua Yi Xue Za Zhi (Taipei) 63: 542-551.

51. Girault C, Richard JC, Chevron V, Tamion F, Pasquis P, et al. (1997) Comparative physiologic effects of noninvasive assist-control and pressure support ventilation in acute hypercapnic respiratory failure. Chest 111: 1639-1648.

52. Laserna E, Barrot E, Beiztegui A, Quintana E, Hernández A, et al. (2003) [Non-invasive ventilation in kyphoscoliosis. A comparison of a volumetric ventilator and a BIPAP support pressure device]. Arch Bronconeumol 39: 13-18.

53. Storre JH, Seuthe B, Fiechter R, Milioglou S, Dreher M, et al. (2006) Average volume-assured pressure support in obesity hypoventilation: A randomized crossover trial. Chest 130: 815-821.

54. Windisch W, Freidel K, Schucher B, Baumann H, Wiebel M, et al. (2003) The Severe Respiratory Insufficiency (SRI) Questionnaire: a specific measure of health-related quality of life in patients receiving home mechanical ventilation. J Clin Epidemiol 56: 752-759.

55. Crisafulli E, Manni G, Kidonias M, Trianni L, Clini EM (2009) Subjective sleep quality during average volume assured pressure support (AVAPS) ventilation in patients with hypercapnic COPD: a physiological pilot study. Lung 187: 299-305.

56. Oscroft NS, Ali M, Gulati A, Davies MG, Quinnell TG, et al. (2010) A randomised crossover trial comparing volume assured and pressure preset noninvasive ventilation in stable hypercapnic COPD. COPD 7: 398-403.

57. Janssens JP, Metzger M, Sforza E (2009) Impact of volume targeting on efficacy of bi-level non-invasive ventilation and sleep in obesity-hypoventilation. Respir Med 103: 165-172.

58. Murphy PB, Davidson C, Hind MD, Simonds A, Williams AJ, et al. (2012) Volume targeted versus pressure support non-invasive ventilation in patients with super obesity and chronic respiratory failure: a randomised controlled trial. Thorax 67: 727-734.

59. Dreher M, Ekkernkamp E, Walterspacher S, Walker D, Schmoor C, et al. (2011) Noninvasive ventilation in COPD: impact of inspiratory pressure levels on sleep quality. Chest 140: 939-945.

60. Bonmarchand G, Chevron V, Ménard JF, Girault C, Moritz-Berthelot F, et al. (1999) Effects of pressure ramp slope values on the work of breathing during pressure support ventilation in restrictive patients. Crit Care Med 27: 715-722.

61. Manning HL, Molinary EJ, Leiter JC (1995) Effect of inspiratory flow rate on respiratory sensation and pattern of breathing. Am J Respir Crit Care Med 151: 751-757.

62. Thille AW, Rodriguez P, Cabello B, Lellouche F, Brochard L (2006) Patient-ventilator asynchrony during assisted mechanical ventilation. Intensive Care Med 32: 1515-1522.

63. Girault C, Briel A, Benichou J, Hellot MF, Dachraoui F, et al. (2009) Interface strategy during noninvasive positive pressure ventilation for hypercapnic acute respiratory failure. Crit Care Med 37: 124-131.

64. Nava S, Navalesi P, Gregoretti C (2009) Interfaces and humidification for noninvasive mechanical ventilation. Respir Care 54: 71-84.

65. Elliott MW (2004) The interface: crucial for successful noninvasive ventilation. Eur Respir J 23: 7-8.

66. Mador MJ, Krauza M, Pervez A, Pierce D, Braun M (2005) Effect of heated humidification on compliance and quality of life in patients with sleep apnea using nasal continuous positive airway pressure. Chest 128: 2151-2158.

67. Saatci E, Miller DM, Stell IM, Lee KC, Moxham J (2004) Dynamic dead space in face masks used with noninvasive ventilators: a lung model study. Eur Respir J 23: 129-135.

68. Schettino GP, Chatmongkolchart S, Hess DR, Kacmarek RM (2003) Position of exhalation port and mask design affect CO2 rebreathing during noninvasive positive pressure ventilation. Crit Care Med 31: 2178-2182.

69. Schönhofer B, Sortor-Leger S (2002) Equipment needs for noninvasive mechanical ventilation. Eur Respir J 20: 1029-1036.

70. Henke KG, Badr MS, Skatrud JB, Dempsey JA (1992) Load compensation and respiratory muscle function during sleep. J Appl Physiol (1985) 72: 1221-1234.

71. Rabec C, Rodenstein D, Leger P, Rouault S, Perrin C, et al. (2011) Ventilator modes and settings during non-invasive ventilation: effects on respiratory events and implications for their identification. Thorax 66: 170-178.

72. Hess DR (2011) Patient-ventilator interaction during noninvasive ventilation. Respir Care 56: 153-165.

73. Berry RB, Chediak A, Brown LK, Finder J, Gozal D, et al. (2010) Best clinical practices for the sleep center adjustment of noninvasive positive pressure ventilation (NPPV) in stable chronic alveolar hypoventilation syndromes. J Clin Sleep Med 6: 491-509.

74. Carrillo A, Ferrer M, Gonzalez-Diaz G, Lopez-Martinez A, Llamas N, et al. (2012) Noninvasive ventilation in acute hypercapnic respiratory failure caused by obesity hypoventilation syndrome and chronic obstructive pulmonary disease. Am J Respir Crit Care Med 186: 1279-1285.

75. Carron M, Freo U, BaHammam AS, Dellweg D, Guarracino F, et al. (2013) Complications of non-invasive ventilation techniques: a comprehensive qualitative review of randomized trials. Br J Anaesth 110: 896-914.

76. Atkeson AD, RoyChoudhury A, Harrington-Moroney G, Shah B, Mitsumoto H, et al. (2011) Patient-ventilator asynchrony with nocturnal noninvasive ventilation in ALS. Neurology 77: 549-555.

77. Vignaux L, Vargas F, Roeseler J, Tassaux D, Thille AW, et al. (2009) Patient-ventilator asynchrony during non-invasive ventilation for acute respiratory failure: a multicenter study. Intensive Care Med 35: 840-846.

78. Jolliet P, Tassaux D (2006) Clinical review: patient-ventilator interaction in chronic obstructive pulmonary disease. Crit Care 10: 236.

79. Adler D, Perrig S, Takahashi H, Espa F, Rodenstein D, et al. (2012) Polysomnography in stable COPD under non-invasive ventilation to reduce patient-ventilator asynchrony and morning breathlessness. Sleep Breath 16: 1081-1090.