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Non-invasive home mechanical ventilation: qualification, initiation, and monitoring

Nieinwazyjna wentylacja w warunkach domowych — kwalifikacja, rozpoczęcie i monitorowanie

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Abstract

Following the introduction of non-invasive positive pressure ventilation (NPPV), the number of patients using home mechanical ventilation has increased substantially and continues to rise worldwide. This is primarily explained by both the effectiveness and comfort that are offered by NPPV in most patients, and particularly in patients with chest wall and neuromuscular diseases. For clinically stable patients the qualification for NPPV largely depends on the presence of complaints or signs of (nocturnal) hypoventilation with accompanying hypercapnia. For patients who are referred by an ICU there are additional prerequisites. In any case, the aims of NPPV should be met and NPPV should be effective. The initiation of NPPV, whether in the clinic or not, should always be tailored to the individual patient. Based on effectiveness, safety, and comfort, the best ventilator has to be chosen. Although with modern interfaces NPPV may be provided continuously, for continuing NPPV over the years, adding manual and/or mechanical cough augmentation techniques is usually mandatory. To control the ongoing effectiveness of NPPV regular monitoring of the patient is essential, and nowadays transcutaneous measurement of CO_2 seems the most reliable and appropriate technique. For trend analysis, downloaded data of modern ventilators may be helpful as well. The ultimate goal of NPPV, to prevent tracheotomy, can only be reached if the patient has continuous access to a centre with expertise in cough augmentation techniques and both nocturnal and diurnal NPPV.

Key words: non-invasive ventilation, qualification, initiation, monitoring

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Streszczenie

Wprowadzenie nieinwazyjnej wentylacji mechanicznej (NWM) zwiększyło liczbę chorych korzystających z wentylacji mechanicznej w domu. Liczba ta stale rośnie na całym świecie. Jest to spowodowane przede wszystkim skutecznością i komfortem NWM u większości chorych, szczególnie u tych ze schorzeniami klatki piersiowej oraz chorobami nerwowo-mięśniowymi. U chorych stabilnych klinicznie kwalifikacja do NWM opiera się na dolegliwościach zgłaszanych w wywiadach i objawach nocnej hipowentylacji z towarzyszącą hiperkapnią. Pacjenci kierowani z oddziału intensywnej opieki medycznej muszą spełniać jeszcze inne kryteria. W każdym przypadku cele NWM powinny być spełnione i NWM powinna być skuteczna. Rozpoczęcie NWM zarówno w klinice, jak i poza nią musi być dostosowane do indywidualnych potrzeb pacjenta. W celu osiągnięcia skuteczności, bezpieczeństwa i komfortu należy wybrać najlepszy respirator. Dzięki wykorzystaniu nowoczesnej maski NWM może być stosowana w sposób ciągły przez lata, chociaż zazwyczaj wymaga wspomagania kaszlu (ręcznego lub mechanicznego). Aby ocenić skuteczność NWM, konieczne jest monitorowanie chorego. Obecnie przezskórny pomiar CO_2 wydaje się najbardziej odpowiednią techniką. Podczas długotrwałych obserwacji uzyskane dane powinny być wprowadzone do pamięci respiratora. Ostateczny cel NWM, którym jest uniknięcie tracheotomii, może zostać osiągnięty, jeśli pacjent ma zapewniony stały dostęp do centrum, które ma doświadczenie w technikach wspomagania kaszlu oraz nocnej i dziennej NWM.

Słowa kluczowe: nieinwazyjna wentylacja, kwalifikacja, rozpoczęcie, monitorowanie

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Introduction

Non-invasive ventilation has been used as early as in the 1800s, but its true advance occurred during the polio epidemics in the first half of the 20th century by means of so-called tank ventilators, including the 'iron lung' [1]. In the late 1950s and 1960s the obvious advantages of invasive positive pressure ventilation led, however, to the virtual substitution of non-invasive ventilation techniques. Interest in non-invasive ventilation began to reappear during the mid 1980s, following the development of a nasal mask for applying continuous positive airway pressure (CPAP) in patients with obstructive sleep apnoea [2]. Soon after, reports began emerging of the positive effects of non-invasive positive pressure ventilation (NPPV) in patients with acute or chronic respiratory failure [3, 4]. From then on things went very fast. Improvements in both ventilators and interfaces have given NPPV a central role nowadays in the care of patients with chronic respiratory failure [5]. In this article three core aspects of NPPV will be discussed: qualification, initiation, and monitoring.

Qualification

In order to qualify for chronic NPPV patients should have a disease that causes both chronic respiratory failure and complaints and/or signs of nocturnal hypoventilation, like restless sleep, nightmares, night sweating, morning headaches, awaking not fit, failure to thrive, difficulty in concentrating, hypersomnolence, orthopnoea, weight loss, and tachypnoea. Recurrent chest infections may be caused by both (nocturnal) hypoventilation and decreased coughing strength. Sitting and supine lung function tests, though helpful in detecting progress of weakness of the respiratory muscles and frequently predictive of nocturnal hypoventilation, are hardly relevant to qualify for NPPV [6]. In addition, patients should have nocturnal hypoventilation or, in a later stage of their disease, chronic respiratory failure, in terms of hypercapnia (PCO_2 > 45 mm Hg or > 6.0 kPa) (Tab. 1). As 70% of patients with nocturnal hypoventilation will develop daytime hypercapnia or secondary complications within one year, it seems prudent to start NPPV once nocturnal hypoventilation is demonstrated [7–9]. In patients with amyotrophic lateral sclerosis (ALS) hypercapnia is not always required to qualify for NPPV, as these patients may have normocapnia at the cost of high work of breathing or are likely to develop (nocturnal) hypercapnia within days or weeks. Even without demonstrable hypercapnia these patients may, however, experience benefit from NPPV.

Patients should also meet the aims of NPPV: maintaining or improving the quality of life with increased independence, exercise tolerance, and better communication; preventing secondary complications like recurrent chest infections and cor pulmonale; and increasing survival [8]. In addition, NPPV should be effective, as shown by an improvement of ventilation, gas exchange, and sleep quality and quantity, and by a reduction of complaints and symptoms [9]. These aims are usually met in patients with neuromuscular diseases and chest wall disorders. In patients with chronic obstructive pulmonary disease (COPD), however, it is still uncertain if these goals can be reached. For COPD patients with chronic respiratory failure in general, several randomized controlled trials have not shown a clear benefit of NPPV, in terms of improved survival or quality of life [10–12]. In the Netherlands, NPPV is thus only offered to COPD patients with chronic respiratory failure as a bridge to transplant [13]. Conceivably, continuation of NPPV in the home setting may be beneficial to a subgroup of COPD patients who used acute NPPV during recurrent hospital admissions for hypercapnic exacerbations [14, 15]. This is at present being investigated in three randomized controlled trials, which are running in France, the UK, and the Netherlands. Until the results of these trials are published, a rather restrictive policy with regard to domiciliary use of NPPV in patients with COPD seems justified.

Table 1. Diseases and level of (nocturnal) hypercapnia for which NPPV may be indicated, depending on the presence or absence of complaints of nocturnal hypoventilation

Tabela 1. Wskazania do NPPV w zależności od schorzenia, stężenia dwutlenku węgla oraz objawów nocnej hipowentylacji

	Without complaints	With complaints
Neuromuscular disease	> 7.0 kPa (52.5 mm Hg)	> 6.0 kPa (45.0 mm Hg)
Motor neuron disease	> 6.0 kPa (45.0 mm Hg)	Irrespective of PCO_2
Chest wall disorder	> 7.0 kPa (52.5 mm Hg)	> 6.0 kPa (45.0 mm Hg)
Obesity hypoventilation syndrome	> 7.5 kPa (56.3 mm Hg)	> 7.5 kPa (56.3 mm Hg)

Finally, there are a number of prerequisites that have to be met, particularly by patients on an intensive care unit (ICU), before home mechanical ventilation (HMV) in general can be considered [16–18]. There should be no other options to treat the respiratory failure, like medication, oxygen therapy, or CPAP. Reversible factors, for example electrolyte disturbances, heart failure, constipation, and medication causing depressed respiratory drive, should be corrected. Patients should be clinically stable as shown by, amongst other things, an inspiratory oxygen fraction ≤ 0.4 and a positive end-expiratory pressure ≤ 10 cm H_2O. If possible, they should have been weaned, but not at the expense of severe hypercapnia. Weaning is not only important to determine the need for long-term ventilation but also to estimate the ventilator-free time, which is important for reasons of safety. Although weaning itself is not a prerequisite for NPPV, the chance of success of transferring patients from invasive ventilation to NPPV seems, to some extent, related to ventilator dependency. NPPV should not be futile, which denotes that patients should have a life expectancy of several months at least. Moreover, patients must be motivated to use NPPV and they should also have a clear perspective for using it for some time, and often indefinitely. Last but not least, the home situation should be safe or adapted to be safe with regard to the physical environment, basic utilities (electricity, heating), alarms, and adequate resources, both financially and with regard to the number of competent caregivers. The latter is usually only considered for patients using invasive ventilation, but patients fully dependent on NPPV may also need more care than the care of family members and lay helpers, also depending on the nature of their underlying disease.

Initiation

Preferably, NPPV should be initiated electively in a quiet atmosphere without pressure on the patient. All equipment and expertise should, ideally, be available to make NPPV both effective and comfortable in the shortest time. For this reason, initiating NPPV in the home setting of a patient may be difficult but not impossible. Randomised controlled trials showed comparable results between in- and outpatient initiation of NPPV in highly selected patients with neuromuscular disease and near normal daytime $PaCO_2$, but without bulbar weakness or cognitive impairment [19, 20]. At present, the initiation of NPPV is usually performed on respiratory wards or respiratory care units.

To be ventilated, a patient needs an interface and a ventilator, which either operates in the volume controlled mode or in the pressure controlled mode. In the volume controlled mode, a fixed volume is delivered during a specified time with any pressure that is necessary to achieve this, regardless of the patient's own ventilation [21]. An inspiratory effort will thus not lead to changes in the delivered volume, but only result in a lower airway pressure. The advantage of this modality is, in the absence of leaks, the fixed delivery of the preset volume, irrespective of the compliance and resistance of the respiratory system. If there is a leak, however, there will be no increase in flow rate to compensate for it and the volume effectively delivered will be diminished.

In the pressure-controlled mode, airflow is delivered by generating a predefined positive pressure in the airways for a given time [21]. Constant analysis of the flow rate and airway pressure determines the flow variations necessary to keep a certain pressure. Consequently, the volume delivered will depend on the interaction between the preset pressure, the inspiratory time, the compliance and resistance of the respiratory system, and the inspiratory efforts of the patient. To overcome this limitation, some new ventilators adjust the pressure level within each cycle or during several cycles to provide a preset target volume, thus combining features of pressure and volume ventilation. This hybrid mode is called volume-targeted pressure ventilation [21]. This mode may improve nocturnal ventilation, particularly in patients with obesity hypoventilation syndrome [22].

For NPPV, most of the time a pressure-controlled ventilator is chosen as this type of ventilator is able to compensate for moderate leaks. There is, however, no proof that pressure-controlled ventilation is in general better than volume-controlled ventilation [9, 23, 24]. For each individual patient the best ventilator has to be chosen based on effectiveness, safety, and comfort. Modern (hybrid) ventilators, instead of offering only one mode of ventilation, have several modes of ventilation. Besides a volume-controlled mode, they may offer a pressure support mode and a pressure-controlled mode, with or without a target volume. In addition, they may have the option of positive end-expiratory pressure (PEEP or EPAP), and they may be used with both an open system, like BiPAP, and a closed system with an expiratory valve. In several machines, two or even three profiles are available for different ventilatory modes and needs. Several of these 'all-in-one' ventilators offer the possibility of a tubing system with one or two ho-

ses. Finally, this new generation of ventilators has the possibility of storing ventilatory data for up to a year and to combine these with data from another source, like a pulse oximeter or capnometer.

Foremost in neuromuscular patients, the settings of the ventilator should be set with the aim to control (nocturnal) ventilation, to reduce CO_2, and preferably to reach normocapnia or an acceptable $PaCO_2$ if normocapnia cannot be reached or is not desired (COPD), and thus reach the aims of NPPV [9, 25]. Simultaneously with striving for normocapnia, the least number of hours on the ventilator should be aimed for. Consequently, nocturnal ventilation needs to be optimally effective before the number of hours on the ventilator during daytime is extended. Furthermore, by aiming for effective ventilation, supplemental oxygen therapy may be reduced or stopped and PEEP-levels can be minimized.

At the same time, the patient has to accept and tolerate an interface [26–28]. For patients with claustrophobia, interfaces with a small nose cap or nasal pillows, which hardly cover the face, may be a solution. Once the patient tolerates the interface and feels comfortable sleeping with it, the effectiveness comes into play. Guidelines or practice standards stating the best interface for each patient do not exist. Finding the best interface is a matter of trial and error. In most patients a nasal mask will do. A nasal mask offers comfort and allows for speech and verbal alarms but may cause a leak through the mouth during sleep, which may reduce the quality of ventilation and sleep [29].

If there is a serious leak through the mouth despite a chinstrap, a nose-mouth (full face) mask may be an option [30]. This is often necessary in patients with a low respiratory compliance, like those with chest wall disorders or obesity hypoventilation syndrome. If both a chinstrap and a full-face mask are not tolerated, humidification may diminish the leak [31].

A hybrid mask combines nasal pillows, which fit into the nares, with a mask that covers the mouth. It may be used in case of (imminent) decubitus of the nasal bridge or for reading or watching television during the daytime while a conventional mask is used during the night.

A total face mask, which is often used for acute NPPV, may be used when other interfaces are not an option. Because of the large dead space and suboptimal fit with a large air leak, a very high flow is usually needed. Some ventilators may not be able to provide such a high flow.

Nowadays, there is such a wide array of interfaces that it is seldom necessary to use a custom mask. The latter may be made of silicon paste or may be ordered through companies that specialise in making these, often very nice but expensive, masks. Whatever mask is chosen, patients should always have a spare mask at home in case a mask becomes damaged. For patients needing more than just nocturnal ventilation, two different masks may be needed to prevent decubitus on the nasal bridge: for example a full face mask during the night and a mask with pillows, which leaves the nasal bridge free, one or more times during the day.

If a patient cannot breathe spontaneously for more than a few hours at a time, diurnal mouthpiece ventilation may become an option [32, 33]. The success of continuous NPPV depends on more than sufficient levels of alveolar ventilation with different interfaces and ventilators, however.

As most patients using NPPV, and particularly those with neuromuscular disease, also have cough impairment due to respiratory muscle weakness, supporting the inspiratory muscles only is not enough. Without cough augmentation techniques many patients will have airways encumbered by secretions, which may result in reduced ventilation and contribute to low ventilation/ /perfusion states, which in turn may cause resorption atelectasis and shunting. Moreover, retained secretions may increase the risk of chest infections and respiratory failure [9, 17, 34]. In addition, the effectiveness of their NPPV may be impaired by the diminishing compliance of the respiratory system, causing insufficient alveolar ventilation [35]. To overcome these potential problems manual and mechanical cough augmentation techniques may be helpful. These techniques include inspiratory muscle aids, such as hyperinflation via air stacking or glossopharyngeal breathing; expiratory muscle aids, such as manually or mechanically assisted cough; and combined inspiratory and expiratory muscle aids [36–38].

Air stacking with a resuscitator balloon or a (volume controlled) ventilator may improve peak cough flows and prevent decreasing the compliance of the respiratory system. In adults air stacking is indicated for a peak cough flow (PCF) < 270 L/ /min or a VC < 1.5 L or < 50% predicted [9, 17]. This technique should be practiced three times a day (reaching five maximal insufflations each time) and more often during chest infections. The effectiveness of air stacking may be improved by combining this technique with manual thoracic or abdominal-thoracic compression. Glossopharyngeal breathing, besides improving peak cough flows, may also be helpful for extending ventilator-free time [36]. In the absence of unconsciousness and

barotrauma risk, mechanical insufflation-exsufflation is recommended when manually assisted coughing and air stacking are no longer effective, particularly during chest infections, thus preventing the need for tracheostomy [17, 39, 40]. The latter should be the ultimate goal of NPPV. To keep patients on NPPV it is usually mandatory to apply diurnal mouthpiece ventilation and cough augmentation techniques in due time.

Monitoring

To monitor the effectiveness, comfort, and side effects of domiciliary NPPV, patients should be visited by a nurse with experience in HMV. In stable patients 1–2 visits per year are usually sufficient. Children and patients with ALS may need more frequent visits, however. Patients should also be given the opportunity to call an experienced nurse during daily office hours and to visit an outpatient clinic for HMV patients [41]. Particularly, patients with continuous NPPV should have continuous (24/7) access to a centre with expertise in both nocturnal and diurnal NPPV [42]. Besides continuous accessibility and availability of experienced caregivers, there should also be continuous technical support from the company that provided the ventilator(s) [43]. Finally, the effectiveness of NPPV may be monitored in the clinic or at home by means of pulse oximetry, capnometry, and ventilator data.

Pulse oximetry has the benefits of simplicity, short set-up time, and short time response (seconds) but the drawbacks of motion artefacts, sensitivity to perfusion, and decreased measurement accuracy at $SaO_2 < 80\%$ [44]. Furthermore, pulse oximetry is unreliable for the detection of hypoventilation, particularly in patients with oxygen supplementation [45]. Though pulse oximetry may be advocated as a screening tool in stable NPPV patients without supplemental oxygen, it is unsuitable for proper evaluation of domiciliary NPPV, even when combined with daytime blood gases [42]. Pulse oximetry may be used, however, to detect hypoxemia caused by airway secretions and imminent chest infections [17, 34].

For evaluation of the adequacy of alveolar ventilation during NPPV, assessment of $PaCO_2$ is essential, and repeated sampling of arterial blood remains the 'gold standard' [26]. However, repeated sampling of arterial or capillary blood will usually cause the patient to awake. The use of an arterial line will generally prevent this but needs specially trained personnel and a special care setting, which often are not available for these patients. Blood for measuring $PaCO_2$ at the end of the night is often sampled after arousal and thus after a short period of daytime ventilation. A normal morning $PaCO_2$ does not, therefore, exclude nocturnal hypoventilation. In addition, blood gases reflect gas exchange at only one point in time and may miss significant changes in CO_2 between measurements. As invasive assessment of $PaCO_2$ seems inappropriate for routine evaluation in stable patients, noninvasive assessment by measuring transcutaneous carbon dioxide tension ($PtcCO_2$) or end-tidal carbon dioxide tension ($PETCO_2$) may be used [42, 44].

$PETCO_2$ is measured with a capnometer, which can store data from a whole night. The relationship between $PETCO_2$ and $PaCO_2$ depends on the physiological dead space and the tidal volume delivered. The former depends on the type and extent of underlying disease; the latter on the ventilatory mode and sleep stage. Non-intentional leaks and varying alveolar ventilation may also influence $PETCO_2$. Consequently, $PETCO_2$ measurements during NPPV are unreliable and should not be used for nocturnal monitoring of these patients [44, 46].

To monitor $PaCO_2$ in patients with NPPV, transcutaneous measurement of CO_2 seems more appropriate [42, 44, 47]. By inducing local hyperaemia of the skin, the capillary blood is 'arterialized' and the permeability of the skin to gas diffusion is improved. The resulting $PtcCO_2$ readings, which are corrected for both skin and patient temperature, are close to $PaCO_2$ values, without inducing local skin reactions or skin burns. Continuous $PtcCO_2$ recordings have shown to be in good agreement with arterial measurements, even with high $PaCO_2$ values [46]. However, all $PtcCO_2$ sensors have a lag time of about two minutes, thus precluding monitoring of swift changes in $PaCO_2$, which might be caused by apnoeas, hypopnoeas, or short-lasting leaks. Other disadvantages of transcutaneous capnometry are the need for periodic recalibration, sensor drift, and high costs compared to $PETCO_2$ measurements. Nonetheless, nocturnal $PtcCO_2$ measurements should be considered as a reliable tool for monitoring $PaCO_2$ in patients using NPPV [44, 48, 49].

Modern ventilators are able to store raw data with respect to airflow and pressure. These data may be downloaded with built-in software and, thereby, provide information with regard to tidal volume, respiratory frequency, minute volume, triggering, leaks, and patient compliance. In addition, these data can be analysed during a certain time span or even cycle by cycle. Some ventilators have the option of reading external polygraphic data on heart rate, SpO_2 and $PETCO_2$ or

$PtcCO_2$. The information thus gathered should be regarded as indicators of trends only, as the validity of these parameters still needs to be determined [44].

Whatever means are used for monitoring patients on NPPV, the monitoring should always be done overnight and preferably also during the daytime, both at rest and during activities. To determine the amount of hours a patient should use the ventilator, monitoring should not only occur during ventilation but also during spontaneous ventilation. Depending on the underlying disease and other factors that might influence the stability of the ventilation, monitoring should be performed 2–4 times per year at least. This may be accomplished by visiting patients, by sending cards with stored data, or by telemedicine [50]. The latter seems attractive, particularly for patients in need of frequent monitoring and for those living in remote areas.

Conclusions

The introduction of NPPV has caused a silent yet highly successful revolution in patients with chronic respiratory failure, particularly in patients with neuromuscular and chest wall disease. For NPPV to be successful, qualification criteria should be strictly followed, initiation should occur electively with availability of expertise and all equipment, carbon dioxide should be reduced, and the ongoing effectiveness of NPPV should be regularly monitored. The ultimate goal of NPPV, to prevent the need for tracheotomy, can only be reached if diurnal mouthpiece ventilation and cough augmentation techniques are applied in due time, meanwhile offering the patient continuous access to a centre with expertise in both nocturnal and diurnal NPPV.

Conflict of interest

None.

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