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Review Article

Update on Noninvasive Management of Ventilatory Pump Failure

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Abstract

This is an update on the noninvasive respiratory management of ventilatory pump failure to prevent CO_2 narcosis, coma, resort to invasive airway tubes, and respiratory causes of morbidity and mortality. Ventilatory support is provided via noninvasive interfaces (NVS) and airways cleared by mechanical in-exsufflation (MIE) via noninvasive interfaces and airway tubes when present. Many patients became up to continuously dependent on noninvasive intermittent positive pressure ventilatory support (CNVS) for 30 years without developing acute respiratory failure or even being hospitalized in many cases. The CNVS users' lives were sustained without resort to tracheostomy tubes. Ventilator unweanable patients extubated to CNVS and MIE to permit definitive noninvasive management is considered. Infants with spinal muscular atrophy type 1 (SMA1) have become CNVS dependent by 3 months of age and are now 20 to 30 years old without tracheostomy tubes despite having 0 ml of vital capacity. A 99% successful extubation rate, including 85% rate per attempt, is reported for unweanable infants with SMA1. Successful extubation has also been reported of 254 of 257 patients unable to pass any ventilator weaning parameters or spontaneous breathing trials. Only two patients with severe upper MND, and one with spinal cord injury and cardiovascular instability, underwent tracheostomies. Thirteen patients in essentially chronic vegetative states were weaned from ventilatory support by using MIE via tracheostomy tubes with no supplemental oxygen and over half were decannulated. Thus, noninvasive interventions, including dependence on up to CNVS and MIE, are alternatives to tracheostomy tubes for patients with ventilatory pump failure.

Keywords: Mechanical Insufflation Exsufflation (MIE); Ventilatory Pump

Introduction

When the ventilatory pump fails, either because of severely weakened respiratory muscles from neuromuscular or some central nervous system conditions, patients require ventilatory support to prevent the toxic effects of hypoxia and hypercapnia. Conventionally, invasive airway tubes are passed along with oxygen for respiratory support. However, since 1953 this has also been accomplished by the use of noninvasive intermittent positive pressure ventilatory support (NVS) and mechanical insufflation exsufflation (MIE) via noninvasive interfaces [1].

In 1957, Dr. Augusta Alba of New York University removed 257 patients from Iron Lungs[1] and put them on portable Bantam™ positive pressure ventilators from which they received full ventilatory support via 15 mm angled mouthpieces (Figure 1). Some of these patients continue to do this to this day, that is, to use CNVS for over 65 years without airway tubes, respiratory complications or hospitalizations, thereby preserving their quality of life. Today, besides mouthpieces, the NVS is delivered via nasal (Figure 2), oronasal, and oral (lip cover) interfaces (Figure 3).

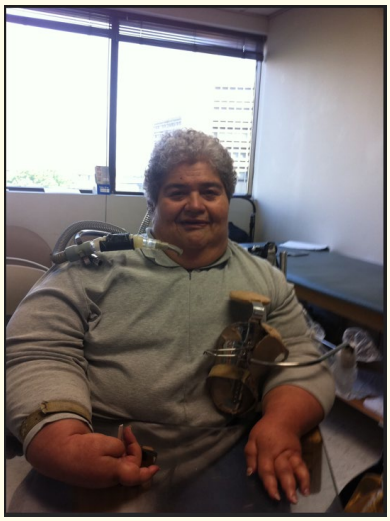

Figure 1: Woman dependent on continuous noninvasive ventilatory support (CNVS) for 70 years from 1953 using mouthpiece NVS for daytime support, lip cover NVS for sleep.

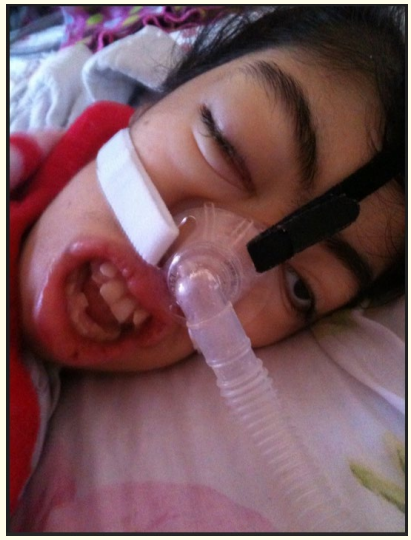

Figure 2: 14 year old girl with SMA type 1 dependent on NVS since age 2, extubated to nasal NVS, no subsequent hospitalizations and today she is 30 years old.

Figure 3: OracleTM interface for noninvasive ventilatory support, Fisher-Paykel Inc., Laguna Hills, CA.

MIE is administered via devices that deliver deep volumes of air, whether via invasive airway tubes or noninvasive mouthpiece, nasal, and oronasal interfaces, to create powerful artificial cough flows to clear the airways and prevent pneumonias for people with ineffective autonomous cough flows. They were on the market from 1953 to 1967 but came off the market when acute on chronic respiratory failure was no longer being prevented by

NVS, and tracheostomies became the convention for tracheostomy mechanical ventilation (TMV) and airway suctioning. No one reported using MIE via the tubes for the next 25 years. Yet, MIE can fully compensate for the denervation and dysfunction of the expiratory muscles that are needed for effective coughing just as NVS compensates for the dysfunction of inspiratory muscles. A sudden mucus plug in a patient with a weak cough can result in a respiratory arrest and encephalopathy.

The term "non-invasive ventilation" or "NIV" has been used for continuous positive airway pressure (CPAP) and bilevel positive airways pressure (BiPAP). While CPAP is almost useless for hypercapnic patients with dysfunctional inspiratory muscles, the settings used for bi-level PAP, too, do not optimally rest respiratory muscles. On the other hand, NVS is used via noninvasive interfaces to optimally rest muscles and provide up to full ventilatory support (NVS) settings. Indeed, volume preset ventilation is delivered at much greater than full support settings to compensate for air leakage in an open system of ventilatory support, and to permit patients to physiologically vary tidal volumes by taking as much of the set volumes as they desire for any particular breath. Volume cycling also facilitates "air stacking" of consecutively delivered volumes of air for lung expansion.

Pathophysiology

Respiration involves three muscle groups: inspiratory, expiratory, and bulbar-innervated. Respiratory failure due to respiratory muscle dysfunction often unnecessarily leads to tracheotomy because CNVS and MIE are not offered. Extubation failures for patients with ventilatory pump failure are usually due to a combination of failure to being too weak to breathe and too weak to cough and effectively clear airway secretions. Patients with upper motor neuron involvement (for example, from ALS) can have spasticity that causes closure of the upper airway, preventing the expulsion of secretions, and undergo tracheotomies. This happens when the hemoglobin saturation (O_2sat) baseline remains below 95% because secretions can't be expelled. However, this does not occur for patients using MIE with myopathies or lower motor neuron diseases (MNDs) [2,3]. The inspiratory and expiratory muscles, but not the bulbar muscles, can be fully compensated for without need for tracheostomy.

Pulmonary function testing evaluates lung and airways diseases but is inadequate for patients with ventilatory pump failure since cough flows, lung volume recruitment, and CO_2 levels are not measured. The treatment for cardiopulmonary diseases may include CPAP or bi-level PAP. However, these are not appropriate or optimal for treating ventilatory pump failure which typically begins with nocturnal ventilatory insufficiency as diaphragm dysfunction causes orthopnea and/or sleep hypoventilation. Hypoventilation can eventually progress to daytime hypoventilation as well. As blood CO_2 levels increase, bicarbonate is retained to counteract the acidosis due to hypercapnia and maintain a more normal blood pH. Hypoxia can also result from hypoventilation and hypercapnia and become severe enough to caused narcosis, coma, and encephalopathy if not properly managed. Providing supplemental oxygen can greatly increase CO_2 levels and lead to ventilatory failure and cardiac arrest. NVS and MIE used instead of O_2 to re-normalize CO_2, O_2, and bicarbonate levels can prevent this and relieve symptoms of ventilatory failure.

Glossopharyngeal breathing (GPB)

Patients with total paralysis of respiratory muscles can ventilate their lungs to normal O_2 and CO_2 levels by GPB provided that they do not have tracheotomies. GPB is performed by the tongue pistoning air past the glottis and into the lungs for up to full tidal volumes. Patients with 0 ml of VC have even awakened from sleep performing GPB to discover that their ventilators were no longer functioning. Had they been using TMV they might have died [4,5]. Some with little or no VC use GPB all day without ventilator use [6-10].

Mechanical insufflation-exsufflation (MIE)

Mechanical insufflation-exsufflation substitutes for an ineffective cough by a positive pressure delivery of air followed by negative pressure expelling the air via oronasal interfaces, mouthpieces, or invasive airway tubes. Since 1953 MIE has only been reported to be most effective when used at pressures of 40 mmHg to -40 mmHg (54.1 cm H_20). These pressures typically generate over 250 L/min of exsufflation flow (MIE-EF) to expel airway debris and prevent pneumonias.

When MIE is used through a tracheotomy or translaryngeal tube, up to maximum device pressures need to be used to compensate for the pressure drop off across narrow airway tubes.

The pressures required are 60 to 70 cm H_2O [11]. MIE is used via translaryngeal and tracheostomy tubes to normalize O_2 saturation and prepare patients for extubation or decannulation [12,13].

MIE is needed and used when patients have cough peak flows under 270 L/m to clear airway secretions and re-normalize O_2 sat levels in room air. It can be used repeatedly, every 20 minutes or more during the 36 hours post-extubation or decannulation to avoid extubation failure, and used in the same manner to treat respiratory infections. MIE is superior to invasive airway suctioning since suction catheters can miss the left mainstem bronchus up to 92% of the time [14-17].

MIE may not provide adequate exsufflation (cough) flows (MIE-EF) for some patients with upper MNDs such as ALS, traumatic brain injury, stroke, etc.. These patients may have collapse of their upper airways [3] and inadequate MIE-EF. With ALS, as the upper airways collapse, MIE-EF decreases. When the MIE-EF are <100 L/min, tracheotomy becomes necessary [2,18], Other than for these cases, other patients with even little to no measurable autonomous cough flows can have MIE-EF greater than 200 L/min. Such flows are effective for clearing secretions, mucus plugs, and preventing cardiopulmonary arrests.

Daytime support

Typically, mouthpiece (Figure 1) and/or nasal NVS (Figure 2) is used during the day for support and to facilitate safe swallowing. Patients with weakened inspiratory muscles typically take more than 40 shallow breaths per minute, which means they only have about a second to gulp down food. To maintain minute ventilation, time for swallowing increases to 10 seconds or more with the use of NVS with delivered volumes of 1 liter or more via a mouth or nose piece.

An alternative to NVS is the intermittent abdominal pressure ventilator (IAPV). A ventilator delivers air, under the user's clothing, into an elastic air sac that compresses the abdomen and moves the diaphragm up. When gravity brings it back down, normal tidal volumes can enter the lungs [37].

When weakening patients become dyspneic when discontinuing nocturnal NVS in the morning, they continue it into daytime hours. This permits weakening patients to transition from nasal to mouthpiece NVS without developing obtundation or CO_2 narcosis (Figure 1). The NVS mouthpieces can be attached to a flexible metal support air on a wheelchair and positioned next to the mouth for easy access. Effective mouthpiece NVS requires some neck and lip movement and strength. Moreover, it is imperative that the soft palate seal off the nasopharynx to prevent excessive air leakage from the nose. Normal reflex opening of the glottis to inhale air is also imperative. Some of these reflex movements can be lost during TMV and need to be relearned for using CNVS after decannulation.[10] The ventilators are set to deliver volumes between 650 and 1,500 ml for daytime assistance/support. This is to allow the patients to vary their tidal volumes to promote lung health and comfort.

Patients use up to CNVS via nasal interfaces when their lips or neck muscles are too weak to grab and retain a mouthpiece or if lack of jaw opening. Although nasal NVS is more often used for nocturnal support, it is a preferable option compared to tracheotomy. When using nasal NVS around-the-clock it is important to vary nasal interfaces day and night to avoid excessive skin pressure causing pressure sores.

Nocturnal support

Nocturnal ventilation is normally begun when the patient has symptoms (Table 1) and evidence of weak respiratory muscles. For nocturnal NVS, one can use different interfaces to the lips, nose, and/or mouth. The recommended prescription is 650 to 1,500 ml or 18 to 25 cm H_2O with a back-up rate of 12 to 14 breaths per minute. The goal is to provide up to full ventilatory support for normal CO_2 levels and to vary tidal volumes, allow active LVR, and optimally rest inspiratory muscles.

Excessive air leakage can be largely avoided by avoiding supplemental oxygen and sedative medications. Passive mechanisms include that the NVS forces the soft palate against the posterior surface of the tongue [20]. Actively, air leakage can be reduced by repeated transient low-level arousals which do not consciously disrupt the patient's sleep [21]. However, the only concern in regard to excessive air leakage is when it disrupts sleep and causes severe O_2 desaturation. In this case, oronasal interfaces are used for sleep NVS (Table 2). Only active circuits with non-ventilated interfaces should be used as discussed below.

Mask type	Definition	Circuit type
Vented interfaces	Have open portals or areas for exhaling air	Passive ventilator circuits for CPAP and bi-level PAP (single limb circuits)
Non-vented interfaces	Do not have open portals for exhaling air	Active circuits, circuits with exhalation valves (two branch circuits), for noninvasive ventilatory support

Table 2: Interfaces and Ventilator Circuits.

During nocturnal NVS, vasodilation and nasal congestion can be caused by dry nasal mucus membranes.[22] A hot water bath humidifier addresses this [22]. The use of decongestants or switching to a lip cover-only interface can also be considered.

Gastroesophageal sphincter integrity is normally to a pressure of 25 cm H_2O, but it can be lower, making patients more vulnerable to abdominal distension. Indwelling gastrostomy tubes can be left open during sleep to prevent this.

NVS may be ineffective for individuals with depressed cognitive function, pulmonary disease that necessitates a high fraction of O_2, uncontrolled seizures, and substance abusers. Diaphragm and phrenic nerve pacing are not warranted or effective for patients with myopathic or lower MNDs. They also cause obstructive events that can either necessitate CPAP, a tracheostomy tube, or simply eliminating them and using NVS.

Lung volume recruitment (LVR)

As VC decreases, total lung volume and pulmonary compliance decrease. This can be eased by performing active LVR. LVR is performed actively by using a manual resuscitator to deliver volumes of air that are held by a closed glottis. This can improve VC, increase cough flows, and prevent or reverse atelectasis [23,24]. The increased lung volumes allow patients to formulate longer sentences with an increase in voice volume. Patients who can perform active LVR can be more easily extubated to CNVS when intubated because they have experience in receiving air volumes via a mouth and/or nose piece. The interfaces used for LVR are the same as for NVS [25].

Specific evaluation for ventilatory pump failure

Symptoms of nocturnal hypoventilation and signs of ventilatory pump failure can be seen in Table 1. Respiratory muscle failure can result in CO_2 narcosis with lethargy and confusion and far worse if not prevented by NVS [26]. Orthopneic patients often have a sitting to supine VC difference over 10%.

Symptomatic patients should be evaluated for blood CO_2 and O_2sat levels. Oximetry, end-tidal CO_2 or transcutaneous CO_2 measurements are adequate. If the patients are symptomatic but the VC is normal in the supine position without significant alterations in O_2sat and CO_2, sleep-disordered breathing (SDB) should be evaluated by polysomnography. However, polysomnographies are not justified for evaluating symptomatic patients with reduced VC because apneas and hypopneas are invariably attributed to central and obstructive events rather than respiratory muscle weakness and this will lead to inappropriate treatment with O_2, CPAP, and bi-level PAP instead of NVS.

When symptoms are unclear, CO_2 and oximetry are monitored during sleep. All symptomatic patients with decreased VC should have a trial of nasal NVS to relieve symptoms and have it prescribed if they benefit from it irrespective of pulmonary parameters. Arterial blood gasometry is rarely if ever warranted [27].

Symptoms	Signs
Morning headache	Orthopnea
Fatigue	Tachypnea
Sleep disturbances	Paradoxical breathing
Hypersomnolence	Hypophonia
	Nasal flaring
	Use of accessory respiratory muscles
	Cyanosis
	Flushing or pallor
	Elevated CO2 levels

Table 1: Symptoms and Signs of Ventilatory Pump Failure.

Long-term management

Patients with breathing problems are often monitored by pulmonary function testing. This evaluates lung and airway function but not muscle function. The forced VC in the sitting

position can be helpful but supine VC and the ratio between the two is far more useful. Capnograph, spirometry, cough peak flow, and oximetry are also needed to evaluate ventilatory pump failure.

Oxygen therapy in patients with NMD may cause respiratory failure, as oxygen will suppress the ventilatory drive and eventually lead to an increase in hypercapnia of 100 mm Hg or more [28,29].

In 2010, clinicians from 22 centers across 18 countries reported a total of 1,623 individuals with spinal muscular atrophy type 1 (SMA1), Duchenne muscular dystrophy (DMD), and amyotrophic lateral sclerosis (ALS) who utilized noninvasive ventilatory support (NVS) to compensate for respiratory muscle dysfunction causing acute respiratory failure. Of the 1623, 760 required NVS continuously (CNVS) for over 3,000 patient years [30].

Intubation and extubation of patients who cannot be weaned from the ventilator

The use of conventional protocols with oxygen therapy and low span bilevel instead of the use of NVS and MIE, for patients with progressive muscle weakness and weak cough, often results in intubation [30], and urging for tracheotomy. Supplemental O_2 is conventionally never turned off. Oxygen supplementation, no matter how minimal, can obscure the reason for otherwise O_2 desaturation. The reasons are hypoventilation, accumulation of secretions in the airways, and the lung disease caused by the latter. To reduce the probability for extubation failure, CO_2 and O_2 sat must be normalized without supplemental O_2 before extubation. This is done by using MIE for effective airway clearing at 60 to 70 cm H_2O pressures via the translaryngeal tubes until O_2 sat is normal and underlying lung pathology cleared. With the administration of oxygen prior to extubation, O_2sat can remain normal despite non-resolution of the underlying pathology and extubation fail.

In 1988, extubation criteria were developed for patients who can not pass ventilator weaning trials or breathe spontaneously. The criteria for extubation to CNVS and MIE include being fully alert and cooperative, unsedated, having no failure of other bodily organs, afebrile, no signs of infection, low to normal blood CO_2 levels, and having O_2sat normal in ambient air for at least 24 hours by using MIE via the tube every hour or so until this is the case. The patient can then be extubated to MIE and to CNVS with typical volumes between 650 and 1500 ml and at physiologic back-up rates and always in ambient air. Upon achieving nasal ventilation, they are then trained to perform mouthpiece ventilation and learn to not allow nasal leakage moving the soft palate to cut off the nasopharynx.

Patients often use mouthpiece NVS and wean from the ventilator by taking fewer and fewer positive pressure ventilations. In the event of O_2 desaturations, insufficient inspiratory pressure should be considered due to air leaks through the interface or tube, hypercapnia, insufficient ventilator settings, or MIE used to correct and maintain sat O_2 >95%. For any O_2 desaturation, inspiratory pressures less than 17-20 cm H2O may signal hypoventilation due to excessive leaks that need to be addressed. MIE should be applied through an oronasal or mouthpiece interface in the event of any O_2 desaturation due to accumulation of bronchial secretions in the airway. Patients and caregivers are taught to administer MIE for O_2 desaturations.

If oral feeding is not safe after extubation, a gastrostomy may need to be performed without intubation or general anesthesia [31-34].

Decannulation

In 1996, 50 ventilator unweanable spinal cord injured patients were reportedly decannulated to CNVS and MIE [35], some had 0 ml of VC and poor bulbar muscle function. We recently decannulated a 26 year old with SMA 1 who was dependent on CNVS since 8 months of age. He has had 0 ml of VC for over 15 years, no bulbar muscle function, and only trace residual eye movement almost since infancy [36]. His MIE-EF via the upper airway were over 350 L/m which is why it can clear his airways and tracheotomy is unnecessary [37]. MIE can need to be used up to every 20 to 30 min until the ostomy is closed and secretions decrease.

Patients without upper MND causing glottis dysfunction and MIE-EF less than 150 L/min may be good candidates for decannulation [38-40]. Whereas TMV typically increases ventilator dependence due to the secretions it causes blocking gas exchange, from deconditioning, and fixed constant tidal volumes [41], decannulation makes it easier to breathe, speak, and swallow. Patients with tracheostomy tubes, even without any ability to breathe free of the ventilator but with VC greater than 250 ml, wean to predominantly sleep NVS after decannulation.

First, after O_2sat baseline is normalized by using MIE via the tubes, fenestrated tubes are placed, and patients practice NVS with the tubes capped (Figures 4, 5). Severe upper MND and generalized brain disease causing upper airway airflow obstruction can be essentially ruled out by observing MIE-EF greater than 200 L/m. Unweanable upper MND patients without lower MND and myopathy patients are then ready for decannulation to CNVS and MIE.

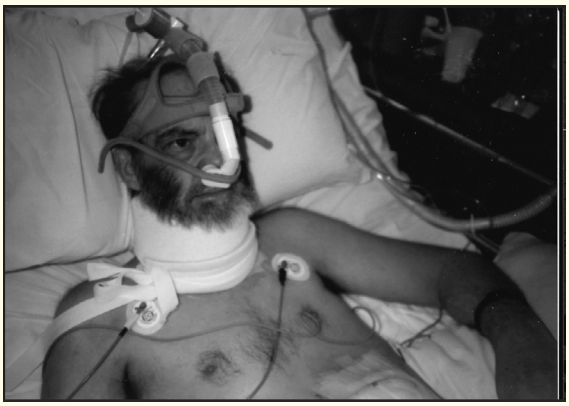

Figure 4: Ventilator unweanable man practicing nasal noninvasive ventilatory support with capped fenestrated tracheostomy tube before decannulation and subsequent dependence on continuous NVS for 14 years.

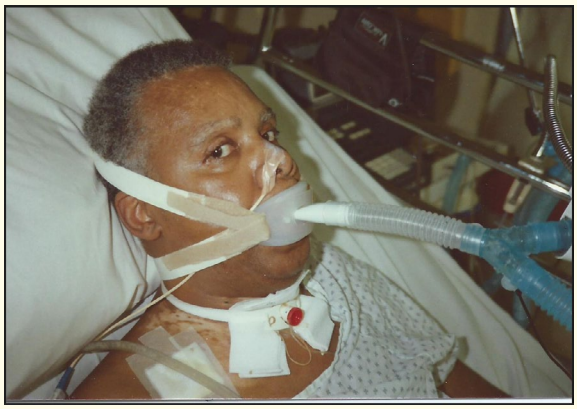

Figure 5: A 42-year-old man with high-level spinal cord injury, with 180 ml of vital capacity, no ventilator-free breathing ability, and the fenestrated tracheostomy tube capped, preparing for decannulation by using mouthpiece NVS and a lip cover for sleep.

Patients with severe upper with brain disease, e.g. brain injury, stroke, cerebral palsy, but without lower MND or myopathy can often only be weaned from ventilator use and use a trach collar after supplemental O_2 is discontinued and MIE used via the tube to clear the airways and normalize O_2sat. Once on trach collars, with the tube capped and MIE used via the upper airways, almost half of the time MIE-EF are adequate for decannulation and to let the ostomy close [42]. High spinal cord injury patients with no ability to ventilate their lungs via a mouthpiece should be considered for phrenic and diaphragmatic pacing.

Oximetry feedback protocol

Tracheostomies should be avoided and reversed, when possible, to permit weaning [43,44]. Oximetry feedback to maintain normal O_2 sat in ambient air using up to CNVS and MIE facilitates this, and to avoid intubation during respiratory tract infections. Patients typically use MIE for all desaturations and extend sleep-only NVS to CNVS as needed.

Manually assisted cough

Cough is manually assisted using an abdominal thrust at glottis opening after a deep inspiration or deep air volume by air stacking if the VC is under 1500 ml [45]. The increased cough flows can prevent pneumonia and respiratory failure [46].

Conclusion and Recommendations

A recent survey of diurnal mouthpiece NVS use had 198 responses with 1 or more patients using sleep plus daytime mouthpiece NVS in each center. There were 116 in Europe, 19 in South Europe, 19 in South America and in the United States, 10 in Australia, 9 in Canada, 9 in Asia, and 16 elsewhere including the Middle-East and Africa. Only 8 centers had over 50 mouthpiece NVS users with six of them cited in www.breatheNVS.com. However, very suboptimal settings of 200 to 400 ml are being used as well as low pressure support levels in most centers [47]. Many are also extending bi-level PAP ventilation into daytime hours at low settings via mouthpieces despite the facts that bi-level, like any pressure preset ventilation and low volume settings, preclude the deep breaths needed for lung expansion and increasing cough flows and voice volumes. Bi-level devices uncomfortably gush air at the patient's throat to compensate for leak, do not have internal batteries for practical daytime use,

and the EPAP is counterproductive and can not be turned off [48]. Pressures of 17 cm or more have been used for hundreds of patients for rest and support since 1957 and preset volumes of 650 to 1800 ml for patients since volume ventilation became available in 1976 [1].

Understanding how aiding and fully compensating for respiratory muscle failure by using up to CNVS and MIE not only sustain life and preserve its quality, but can also save up to $440,000 per year in nursing care as well as literally millions of dollars in charges for tracheotomies and prolonged futile critical care weaning attempts. With over $5 trillion being paid in the U.S. for medical care this year, this may be an appropriate time for a paradigm shift to more humane noninvasive management.

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