

## Noninvasive Management of Ventilatory Pump Failure

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### ABSTRACT

#### Introduction and Objectives

Until resorting to tracheotomy became popular in the late 1960s, all ventilator dependent people were managed noninvasively, that is, by body ventilators until 1953 then subsequently, by noninvasive intermittent positive pressure ventilatory support (NVS) delivered via 15 mm angled mouthpieces during the daytime and mouthpieces with lip cover phalange for sleep [1]. When patients developed respiratory tract infections that resulted in pneumonia and acute on chronic respiratory failure (ARF), underwent translaryngeal intubation for invasive ventilatory support and remained ventilator unweanable, they underwent tracheotomies for Tracheostomy Mechanical Ventilation (TMV). Subsequently, considerable morbidity and mortality have been reportedly due to the tubes themselves. Since no one who does not absolutely need a tracheostomy tube wants one [2], the purpose of this paper is to describe how and when tracheotomies can be avoided in favor of strictly noninvasive management.

We will also describe why patients with Ventilatory Pump Failure (VPF) who require NVS and whose Cough Peak Flows (CPF) cannot exceed 270 to 300 L/m require access to Mechanical Insufflation-Exsufflation (MIE) to clear airway secretions as needed [3]. Also, while “noninvasive ventilation” or “NIV” is often reportedly being used for these patients, NIV has become synonymous with Continuous Positive Airway Pressure (CPAP) and bi-level PAP used at grossly inadequate settings (“spans” or “drive pressures”) for ventilatory support. Patients confined to using only NIV have never been described to be dependent on continuous NVS (CNVS).

#### PATHOPHYSIOLOGY OF VENTILATORY PUMP FAILURE

Alveolar ventilation and airway clearance involve inspiratory muscles, expiratory muscles, and bulbar-innervated muscles. The inspiratory and expiratory muscles make up the ventilatory pump. In the absence of inspiratory muscle function, expiratory (abdominal) muscles can compress the abdomen to raise the diaphragm so that gravity subsequently descends the diaphragm and ventilates the lungs. This is essentially the mechanism of action of the Intermittent Abdominal Pressure Ventilator (IAPV) [4,5]. Ventilatory pump failure can result from insidious elevation of blood CO<sub>2</sub> levels and hypoventilation or it can be acute and triggered by an acute respiratory tract infection and airway congestion due to ineffective cough flows and lead to pneumonia (URI-pneumonia) and possibly Acute Respiratory Failure (ARF). Patients with patent upper airways can use NVS for ventilatory support and MIE for

airway clearance whereas patients with diminished upper airway patency associated with upper motor neuron disease (MND) often cannot. Thus, patients with myopathic or lower MND maintain upper airway patency sufficiently for effective expulsion of airway secretions by MIE and should not require tracheostomy tubes for ventilatory support or airway clearance whereas upper MND patients often do. Upper MND stridor and airway instability can also render NVS ineffective as well as diminish MIE exsufflation flows (MIE-EF) to necessitate tracheotomy.

Ventilatory pump failure initially manifests itself as nocturnal hypoventilation, often associated with respiratory orthopnea caused by severe diaphragm dysfunction. Hypercapnia eventually progresses into daytime hours. As blood CO<sub>2</sub> increases, the kidneys retain bicarbonate to compensate for the hypercapnia and maintain normal blood pH. Hypoventilation also causes hypoxia. However, if supplemental oxygen is given, respiratory drive decreases and this can exacerbate ventilatory insufficiency and CO<sub>2</sub> retention and result in ventilatory arrest [6].

## CLINICAL PRACTICE

### Assessment

Patients with VPF develop symptoms that can include morning headache, fatigue, sleep disturbances, and hypersomnolence. Hypercapnia develops as a result of hypoventilation [7]. Ventilatory insufficiency can be identified by orthopnea, tachypnea, paradoxical breathing, hypophonia, nasal flaring, use of accessory respiratory muscles, cyanosis, flushing or pallor, elevated CO<sub>2</sub> levels, and airway congestion. CO<sub>2</sub> narcosis may be present with lethargy and confusion. Patients' orthopnea is typically identified by VC difference of 30% greater when sitting than when supine. For infants, VPF is also manifested by paradoxical breathing along with sleep flushing, perspiration, and frequent arousals.

Patients suspected of VPF are conventionally sent for Pulmonary Function Testing (PFT), However, PFTs are designed to assess lung and airways diseases but not for muscle dysfunction for which forced expiratory flows are unnecessary. Diffusion studies and plethysmography are also unnecessary. Spirometry needs to be done in sitting, supine, and possibly side lying positions as well as with thoracoabdominal orthotics on and off. Use of thoracolumbar bracing can increase VC

while ill-fitting ones can reduce it. While inspiratory and expiratory muscles can be substituted for without resort to tracheotomy, upper motor neuron bulbar-innervated muscle dysfunction cannot. These patients are also typically sent for polysomnograms. Polysomnograms are programmed to interpret apneas and hypopneas as being due to central or obstructive events but not muscle weakness. The patients' apneas and hypopneas are typically titrated away at bi-level PAP levels much lower than those needed for full respiratory muscle rest or ventilatory support. In fact, apnea-hypopnea indices are typically normalized without normalization of CO<sub>2</sub> levels and the patients remain symptomatic for hypercapnia.

Instead of typical PFTs and polysomnographies, besides spirometry as noted above, Cough Peak Flow (CPF) measurements, assisted CPF measurements, O<sub>2</sub> saturation, and end-tidal CO<sub>2</sub> levels need to be measured [8]. Arterial blood gases are typically unnecessary for NMD patients [9] because oximetry and capnography can provide the needed information. All symptomatic patients with diminished pulmonary function deserve a trial of NVS. However, if pre-treatment symptoms are not obvious, CO<sub>2</sub> and O<sub>2</sub> saturation levels are monitored during sleep to further assess need for NVS.

## APPROACHES TO SUPPORT A FAILING VENTILATORY PUMP: NONINVASIVE VENTILATORY SUPPORT

Symptomatic patients with increased CO<sub>2</sub> and O<sub>2</sub> desaturations during sleep are prescribed NVS to relieve symptoms and normalize blood gases. Any supplemental O<sub>2</sub> discontinued since patients with normal lung tissues can maintain normal blood gases by NVS and MIE alone. On the other hand, if a patient has good muscle strength, normal supine VC, no significant O<sub>2</sub> desaturation or CO<sub>2</sub> retention, then a polysomnogram is used to evaluate for sleep disordered breathing.

### Nocturnal Support

Patients often start nocturnal NVS for relief of symptoms. Lip cover, nasal, and oronasal interfaces can be used to deliver the typical volumes of 800 to 1,500 ml or pressures of 18 to 25 cm H<sub>2</sub>O for NVS with a physiologic back-up rate of 12 to 14 breaths per minute. The large volumes compensate for air leakage and permits users to physiologically vary tidal volumes; it allows active Lung Volume Recruitment (LVR); and it

rests inspiratory muscles [10]. Excessive air leakage that can make NVS ineffective is generally precluded by avoiding supplemental O<sub>2</sub> and sedative medications. An unsedated ventilatory drive reflexively prevents excessive leakage during sleep by repeated transient low-level arousals which do not consciously disrupt the patient's sleep [11]. A passive mechanism that prevents excessive leakage is the nasal NVS that passes via the nose and propels the soft palate against the posterior surface of the tongue [12]. Occasionally air leakage is can be excessive and disrupt sleep and cause prolonged severe O<sub>2</sub> desaturation. The nasal and oronasal interfaces for NVS can be vented or non-vented. Vented interfaces have open portals or areas that allow interface leakage. These vented interfaces are used with "passive" ventilator circuits and deliver CPAP and bi-level PAP. Non-vented interfaces do not have open portals or leak areas and are used with "active" circuits, circuits with exhalation valves. Active circuits should only be used with non-vented interfaces, or vented interfaces with all open areas covered or blocked. Humidity is another factor to consider when using nocturnal NVS. Dry nasal mucous membranes can cause vasodilation and nasal congestion. In the case of nasal NVS, the unidirectional airflow, with expiration through the mouth, can cause loss of humidity and increase airflow resistance [13] One of the ways to alleviate this is to use a hot water bath humidifier [13]. Other options include the use of decongestants. Normal gastroesophageal sphincter pressure is 25 cm H<sub>2</sub>O but is often lower in patients with NMD, thus there is a predisposition to bloating and abdominal distension, even without NVS. The addition of NVS can exacerbate the abdominal distension and, at times, require placement of an indwelling gastrostomy tube to burp out the air. NVS is contraindicated in conditions which prevent reliable access to the NVS interface such as with depressed cognitive function, some orthopedic conditions, pulmonary disease that necessitates a high fraction of O<sub>2</sub>, uncontrolled seizures, and substance abuse. Diaphragm and phrenic nerve pacing are never indicated in patients with VPF other than for certain patients with high level spinal cord injury [14] Pacing requires presence of a tracheostomy tube or CPAP administration because of the obstructive apneas it causes whereas NVS ventilates the lungs and prevents obstructive apneas as well.

### Daytime Noninvasive Ventilatory Support and Ancillary Techniques

The most important interfaces for daytime NVS are 15 and 22 mm mouthpieces (Figure 1). Often mouthpiece NVS is initially used for daytime support to aid with eating [15]. Patients with increasing inspiratory muscle weakness can become tachypneic with breathing rates over 40 breaths per minute which means they only have about 1 second for swallowing food. With the use of NVS and breaths of 1 liter or more to maintain minute ventilation, time for swallowing increases to 10 seconds or more [15].

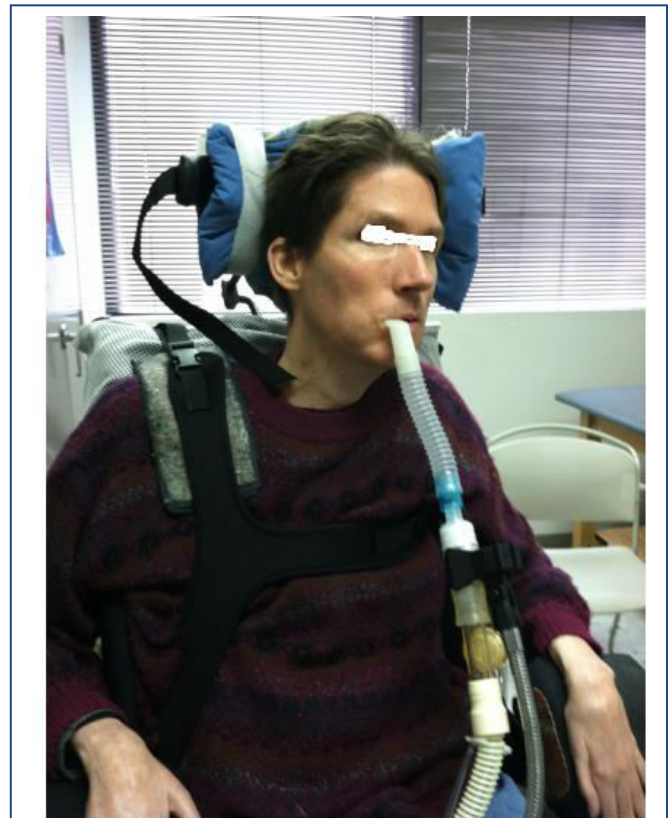


Figure 1: 48 year old with Duchenne muscular dystrophy, dependent on Continuous Noninvasive Ventilatory Support (CNVS) since age 23, using mouthpiece NVS for daytime support, nasal NVS for sleep.

Patients using sleep NVS who become dyspneic when disconnected from it in the morning, tend to continue nasal NVS into daytime hours. At this point patients are transitioned from nasal to mouthpiece NVS. NVS mouthpieces can also be mounted onto a wheelchair and the mouthpiece fixed adjacent to the mouth for easy access (Figure 1). However, some neck and lip function are required to grab the mouthpiece for NVS.

In addition, the soft palate needs to seal off the nasopharynx to prevent air leak from the nose and reflex opening of the glottis. To breathe in the air sometimes has to be relearned for patients who had been intubated [16]. As for sleep NVS, 800 to 1,500 ml delivered volumes are also used for daytime support and for the same reasons, to compensate for leakage to permit active LVF (Lung Volume Function), and for deep breaths to increase CPF and speak longer and louder. For those whose lips or neck muscles are too weak or jaw opening insufficient to grab a mouthpiece, nasal prongs or other nasal interfaces are used for daytime NVS (Figure 2). Nasal NVS is always a preferable option to resorting tracheotomy. When using nasal NVS around-the-clock it is important to alternate two or more nasal interfaces for daytime and sleep use. Often prongs are preferred for daytime use. The Intermittent Abdominal Pressure Ventilator (IAPV) is an effective ventilator for daytime ventilatory support for many patients and is preferred over using facial interfaces during daytime hours. It consists of a girdle or corset worn under the clothing with a cylindrical elastic air sac that, when inflated by a portable ventilator, raises the diaphragm so that gravity can bring it back down to cause air to enter the upper airways. The IAPV can augment tidal volumes by 300 ml to 1,200 ml and is a cosmetic and comfortable method for daytime ventilatory support [4].

Glossopharyngeal Breathing (GPB) involves the movement of tongue and glottis to piston boluses of air into the lungs. Most breaths of 500 ml are accomplished by about 7 such boluses. The pistoning can supplement autonomous breathing or completely substitute for it for patients with little or no Vital Capacity (VC). Its progress can be measured by spirometry. The GPB or “frog breathing” can assist both inspiratory and expiratory functions [17,18]. It can often permit patients with little or no VC to breathe free of ventilatory support up to all day. Patients who master it do not need to worry about ventilator failure [19-21]. Patients with good bulbar-innervated muscle function are the best candidates to master GPB [16,22].

Lung Volume Recruitment (LVR) can be active or passive. Pulmonary compliance decreases with inability to breathe deeply and expand the lungsto predicted normal lung volumes. Lung expansion can be measured by inspiratory capacity which

correlates inversely with extent of chest wall contractures and lung and chest wall restriction. This is treated and reversed by lung volume recruitment (LVR) [23]. The LVR can maintain lung and chest wall compliance, promote lung and chest wall growth for children, and maximize lung inflation and volumes [24].

Active LVR can be achieved by GPB or by air stacking of consecutively delivered volumes of air delivered by volume preset ventilation or manual resuscitator. LVR can increase VC, increase CPF, and reduce atelectasis [25,26]. It can also permit patients to speak louder and longer phrases. If any patient using NVS is able to perform active LVR or “air stacking”, transitioning from extubation to CNVS is easier. The LVR can be provided via mouthpiece, lip cover, or nasal/oronasal interfaces.



Figure 2: 24 year old woman with spinal muscular atrophy type 1, NVS dependent since age 2, successfully extubated for the 11th consecutive time to Continuous Noninvasive Ventilatory Support (CNVS) at age 14 with no hospitalizations since then.

For patients who cannot air stack because they cannot close their glottises, passive LVR can be performed by using a manual resuscitator with blocked exhalation valve to prevent exhalation until maximum lung volumes are attained. Adequate

delivery can be noted by feeling the resistance of lung recoil while squeezing the manual resuscitator to inflate the lungs. Passive LVR can also be provided by mechanical insufflation, which is the insufflation phase of MIE, to pressures of 55-60 cm H<sub>2</sub>O or more. Infants and small children cannot air stack or cooperate with active LVR. However, passive LVR can be provided for them by placing them on sleep nasal NVS, timing manual resuscitator insufflations to their inhalation via oronasal interface, or having them trigger MIE by using the Cough-Track™ Auto-trigger mode on a Cough Assist™. Nocturnal NVS is indicated for all infants with paradoxical chest wall movement to reverse the paradoxing and to prevent/reverse pectus excavatum and promote lung growth. This cannot be accomplished by bi-level PAP at the conventionally prescribed drive pressures (spans).

Progress in mastering GPB and active LVR is monitored by spirometry. For patients with weak lips, air stacking can be provided via nasal/ lip cover or oronasal interfaces. Bulbar innervated muscle function can be assessed by the determination of maximum insufflation capacity which is the maximum amount of air that can be air stacked and then exhaled into a spirometer, minus the VC. MIC (Maximum Insufflation Capacity) and VC difference correlates with bulbar-innervated muscle function.

## APPROACHES TO SUPPORT A FAILING VENTILATORY PUMP: ASSISTED COUGHING AND MECHANICAL INSUFFLATION- EXSUFFLATION

### Assisted Coughing

**Manually assisted coughing:** Manually assisted coughing is performed by taking a deep inspiration to at least 1500 ml or air stacking maximally then having an abdominal thrust timed to glottis opening for coughing. A study showed that with manually assisted coughing subjects' CPF increase from 150±120L/min to CPF of 255±100 L/min [26]. Increased flow scan effectively prevent pneumonias and ARF from URI's [27].

Patients with the ability to air stack but unable to achieve a CPF of >160 L/min should be evaluated for upper airway obstruction with laryngoscopy to assess for reversible lesions. If patients are unable to air stack, likely the inability to close the glottis, they are still able to have manually assisted coughing with abdominal thrusts after deep inspirations. This may increase CPF over 160 L/min as well.

**Mechanical insufflation-exsufflation (MIE):** Mechanical insufflation-exsufflation is essentially mechanically assisted coughing. It is usually used via oronasal interfaces or simple mouthpieces. In these cases, pressures of 40 mm Hg to -40 mm Hg (54.1 cm H<sub>2</sub>O), that is, insufflations to clinically full chest expansion to deep exsufflation to clinically complete chest emptying are optimal. MIE is also used via translaryngeal and tracheostomy tubes at pressures of 60 to 70 cm H<sub>2</sub>O due to the severe pressure drop-off and decreased air flows across the tubes [28].

MIE is used to prevent URI-pneumonias as well as to prepare patients for extubation or decannulation if they get pneumonia and develop ARF anyway [29,30]. About 20% of the time MIE-Exsufflation Flows (MIE-EF) can be increased by applying a manual thrust during the exsufflation phase. MIE treatments typically last until secretions are no longer expelled and secretion-related O<sub>2</sub> desaturations are resolved. During chest infections or 36 hours post-extubation or decannulation, MIE can be used as often as every 20 to 30 minutes around the clock to avert extubation failure.

In comparing MIE to upper airway or invasive airway suctioning, MIE has several advantages. The left main stem bronchus is missed with routine airway suction about 90% of the time [31], while MIE-EF can clear both left and right airways without the discomfort or potential airway trauma from suctioning. This is also a reason why patients prefer MIE over airway suctioning [32]. Effective clearance of airway secretions with MIE improves VC, pulmonary flow rates, and O<sub>2</sub> saturation. In 67 patients with 'obstructive dyspnea', increases in VC of 15% to 40% were reported. In patient with NMD, an increase of 55% in VC was noted without adverse effects [33]. Furthermore, in patients with NMD during chest infections, an improvement of 15% to 400% in VC and normalization of O<sub>2</sub> saturation were reported with the use of MIE [34].

For patients with central nervous system (CNS) or upper MND such as some ALS patients, hypertonicity collapses the upper airways too much for effective MIE-EF [35,36]. When the MIE-EF are <100 L/min, tracheotomy is typically necessary [36]. A MIE-EF of >200 L/min usually very effectively clears secretions and is achievable by all with NMD except for cases of

advance ALS which is why tracheostomy tubes are generally only required by ALS patients.

**LONG-TERM DOMICILIARY PREVENTION OF RESPIRATORY COMPLICATIONS BY THE OXIMETRY FEEDBACK PROTOCOL**

Oximetry feedback can be used essentially all day during URIs or even many times daily for patients who have a tendency to aspirate upper airway secretions to prevent URI-pneumonias and ARF. It is also important along with CNVS for successful extubations and decannulations. Oximetry feedback monitors the normalization of CO<sub>2</sub> and O<sub>2</sub> sat levels by using NVS and MIE. An O<sub>2</sub> saturation alarm can be set to 94% so that with decreases in O<sub>2</sub> sat, the patient can use NVS and/or MIE to clear airway secretions to re-normalize O<sub>2</sub> sat levels. The alarm can also signal the patient to take in deeper air volumes with or without NVS to normalize alveolar ventilation. O<sub>2</sub> desaturation below 95% during URIs is typically due to bronchial mucous plugs which can lead to atelectasis, pneumonia, and collapsed lung, all of which can be prevented by oximetry feedback from use of NVS and, especially, MIE effectively.

**CRITICAL CARE MANAGEMENT**

Conventional management with O<sub>2</sub> and low span bilevel PAP instead of NVS and MIE often results in CO<sub>2</sub> narcosis and respiratory arrest. Once patients are intubated, ventilator weaning parameters and spontaneous breathing trials must usually be passed before any attempt is made at extubation

and the extubations are typically transferred to supplemental O<sub>2</sub> and low span bi-level PAP. However, ventilator unweanable patients can be extubated to full CNVS and MIE rather than weaned to be extubated.

(Table 1) demonstrates the criteria for extubating ventilator unweanable patients developed in 1988 [7]. The O<sub>2</sub> sat must be normal in ambient air. Even a FiO<sub>2</sub> of 25% can prevent an oximeter from signaling airway secretion congestion and marked hypercapnia. Chances of extubation success are decreased if CO<sub>2</sub> and ambient air O<sub>2</sub> sat are not normal before extubation, if airway secretions are not effectively expelled by using MIE via the translaryngeal tube, and if the lung pathology is not corrected prior to extubation by using MIE hourly via the translaryngeal tube. An abnormal ambient air O<sub>2</sub> sat points to these whereas with O<sub>2</sub> administration the O<sub>2</sub> sat can remain normal in the presence of these abnormalities and lead to extubation failing. For this reason, we rarely extubate anyone whose O<sub>2</sub> sat in ambient air less than 95%.

After the criteria are met, any orogastric/nasogastric tubes are removed to facilitate post-extubation nasal NVS. The patient is then extubated directly to CNVS on assist/control mode with preset pressures of about 20 cm H<sub>2</sub>O or volumes of 800-1500 mL at a rate of 10-14 per minute in ambient air. If the patient was using NVS prior to intubation he/she is extubated to the same settings and interfaces. Once they achieve ventilation via nasal interface, mouthpiece NVS is taught then air stacking (Figure 2).

Table 1: Criteria for Extubation of Ventilator Unweanable Patients
Must be fully alert and cooperative
O <sub>2</sub> and sedative medications discontinued
Failure of respiratory function alone with other organs very functional
Afebrile
Normal WBC
Chest X-rays indicating resolving abnormalities
Co <sub>2</sub> tension less than 44 mm HG or end-tidal CO <sub>2</sub> normal
Oxyhemoglobin sat of 95% or higher for at least 12 hours
With translaryngeal tube cuff deflation adequate air leakage through the vocal cords for vocalization

Patients keep 15 mm angled mouthpieces within easy access to their mouths and wean themselves by taking fewer and fewer intermittent positive pressure ventilations. Diurnal nasal NVS is used for those who cannot grab and use a mouthpiece properly. If O<sub>2</sub> sat decreases to less than 95%, ventilator positive inspiratory pressures, interface or tubing air leakage,

CO<sub>2</sub> retention, ventilator settings, and MIE can be considered to reverse the desaturation. Low ventilator positive inspiratory pressures (PIP) indicate air leakage or inadequate settings. MIE is applied via oronasal interfaces at 50 to 60 cm H<sub>2</sub>O to correct any decreases in O<sub>2</sub> sat due to airway mucus. This is done by the patients' family or care providers up to every 20

minutes post-extubation and for all O<sub>2</sub> desaturations below 95%. This is the ideal setting for family members to learn MIE and NVS management as hospital staff will not use MIE sufficiently to facilitate successful extubation.

If post-extubation oral intake is unsafe or inadequate, a gastrostomy tube is needed. This is typically done by radiographically inserted gastrostomy [37], by either open gastrostomy by general surgery [38], or by a percutaneous endoscopic gastrostomy with trochanter passed via an orifice in an oronasal interface being used for NVS during the procedure [39]. These methods permit gastrostomy tube placement without intubation or general anesthesia.

### DECANNULATION

In 1996, decannulation for ventilator dependent spinal cord injury patients and 50 unweanable neuromuscular disease patients was reported [40]. Decannulation is recommended for any patient whose bulbar-innervated musculature is sufficient such that saliva aspiration does not cause a continuous decrease in baseline O<sub>2</sub> sat below 95% and MIE-EF with the upper airway with the ostomy covered is over 150L/min [35] (Figure 3). Patients are decannulated to CNVS in ambient air as their care providers use MIE up to every 20 to 30 minutes to maintain O<sub>2</sub> sat greater than 94% for the first 36 hours following decannulation. Patients with tracheostomy tubes who have no ventilator free breathing ability but who have VCs of 250 mL or greater invariably wean to less than CNVS after decannulation. Their VCs increase, and many wean to nocturnal-only NVS within 3 weeks of decannulation. Only patients with severe NMD glottis dysfunction that results in O<sub>2</sub> desat are poor candidates for decannulation [18]. One study showed decannulation patients prefer CNVS to CTMV (Continuous Tracheostomy Mechanical Ventilation) for all issues studied [2].

Reasons why TVS increases ventilator dependence include tube triggered airway secretions that block respiratory exchange membranes, bypassing upper airway afferents, and respiratory muscle deconditioning [41]. Removal of the tube facilitates speech and swallowing and VCs tend to improve. Phrenic and diaphragm pacing should be limited only to high level spinal cord injury patients with little or no measurable VC and no ability to rotate neck to grab a mouthpiece.



Figure 3: 42 year old high level spinal cord injured patient with 180 ml of vital capacity and no ventilator free breathing ability preparing for decannulation by using sleep lipseal mouthpiece noninvasive ventilatory support with the fenestrated tracheostomy tube capped.

### Outcomes

An April 2010 consensus of clinicians with 760 CNVS dependent patients with NMD noted that patients with DMD live 10 years longer when using CNVS rather than CTMV [8,42]. In another study, patients preferred CNVS for safety, convenience, swallowing, speech, appearance, and comfort [2]. For patients with DMD, 101 became CNVS-dependent for  $7.4 \pm 6.1$  years to a mean of  $30.1 \pm 6.1$  years of age with 56/101 still alive. Twenty-six of the original 101 patients became CNVS without hospitalization or developing ARF [43]. At least 80 intubated DMD patients who could not pass spontaneous breathing trials before or after extubation have now been successfully extubated to CNVS and MIE. CNVS is also an alternative to TMV in the perioperative management of children with flaccid neuromuscular scoliosis who have less than 40% of predicted normal VC [44]. Thus, CNVS provides more favorable results in terms of morbidity and mortality compared to TMV and tracheostomies should be avoided or reversed when possible [45,46].

### CONCLUSIONS

Therefore, ARF can develop from hypoventilation or from inadequate CPF during chest infections for which NVS can be used to re-normalize CO<sub>2</sub>, O<sub>2</sub> sat, bicarbonate levels and reverse chronic alveolar hypoventilation as well as to permit the extubation of ventilator unweanable patients without resorting to tracheotomy [4,47,48]. Thus, in our experience,

only VPF patients with amyotrophic lateral sclerosis/upper MND may eventually require tracheostomy tubes for ventilatory support and airway clearance. This occurs when the oxyhemoglobin saturation baseline decreases below 95% due to inability to expel saliva and airway debris [35,49-51]. Whereas inspiratory and expiratory muscle failure can be substituted for without resort to tracheotomy, upper motor neuron bulbar-innervated muscle dysfunction may not be. Whereas it may be argued that the conclusions of this review are “not evidence-based,” it is obvious that it is not possible to do a placebo controlled study when the intervention being used replaces the function of a vital organ or the vital organ itself. No one with airway secretions during a URI who cannot cough or have cough flows provided for them would survive more than a few hours just as no one with a VC of 0 ml would survive more than a few minutes without full ventilatory support, whether invasive or noninvasive [52]. While it has not been demonstrated other than by historical controls that noninvasive management prolongs life more than invasive management [53] It has also not been demonstrated that invasive management prolongs survival longer than noninvasive management. However, noninvasive management should always be favored to preserve quality of life until it is demonstrated to be inferior to invasive management. Therefore, it is time for a paradigm shift from the latter to the former.

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