NONINVASIVE RESPIRATORY MANAGEMENT AND DIAPHRAGM AND ELECTROPHRENIC PACING IN NEUROMUSCULAR DISEASE AND SPINAL CORD INJURY

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ABSTRACT: The purpose of this monograph is to describe noninvasive management of respiratory muscle weakness/paralysis for patients with neuromuscular disease (NMD) and spinal cord injury (SCI). Noninvasive ventilation (NIV) assists and supports inspiratory muscles, whereas mechanically assisted coughing (MAC) simulates an effective cough. Long-term outcomes will be reviewed as well as the use of NIV, MAC, and electrophrenic pacing (EPP) and diaphragm pacing (DP) to facilitate extubation and decannulation. Although EPP and DP can facilitate decannulation and maintain alveolar ventilation for high-level SCI patients when they cannot use NIV because of lack of access to oral interfaces, there is no evidence that they have any place in the management of NMD.

RESPIRATORY EVALUATION AND MANAGEMENT PARADIGMS

Respiratory impairment can result from either lung or airways disease, in which case pulmonary function testing (PFT) and supplemental oxygen may be appropriate. This approach is inappropriate, however, for patients with primarily ventilatory impairment. The former is characterized by hypoxia in the presence of eucapnia or hypocapnia until hypercapnia signals end-stage respiratory failure. The latter is characterized by hypoventilation-induced hypercapnia and hypoxia with risk of intercurrent episodes of acute respiratory failure (ARF) due principally to an ineffective cough. Unless the latter is distinguished from the former, unnecessary morbidity, airway cannulation, electrophrenic pacing (EPP) or diaphragm pacing (DP), and tracheotomy will result.

There are 3 respiratory muscle groups: inspiratory muscles; expiratory (predominantly abdominal and chest wall) muscles, mainly for coughing; and the bulbar-innervated muscles. Although the inspiratory and expiratory muscles can be completely supported noninvasively, there are no effective noninvasive measures to counter aspiration from bulbar dysfunction. Thus, the only indication for tracheotomy in an alert neuromuscular disease (NMD) or spinal cord injury (SCI) patient is when aspiration results in persistent oxyhemoglobin saturation (SpO₂) of <95% despite noninvasive ventilation (NIV) and mechanically assisted coughing (MAC). Fortunately, other than for advanced bulbar amyotrophic lateral sclerosis (ALS) patients, those with most other NMDs and SCI rarely have bulbar dysfunction to the extent that speech, deglutition, and adequate airway protection are lost, and therefore they can be managed noninvasively.

Intact phrenic nerves, their anterior horn cells, and diaphragm are required for effective EPP/DP. Although EPP/DP can ventilate the lungs of patients with intact anterior horn cells, peripheral nerves, and diaphragms, as in many high-level SCI patients, EPP/DP is not indicated for ALS or other NMDs.

RESPIRATORY MUSCLE AIDS

Inspiratory and expiratory muscle aids include NIV and MAC for NMDs, and NIV, MAC, and EPP/DP for SCI patients. These are techniques that involve the manual or mechanical application of forces to the body, intermittent pressure changes to the airways, or electric current to the phrenic nerves or diaphragm to assist in or substitute for respiratory muscle function.

Inspiratory Muscle Aids. Inspiration can be supported by negative pressure ventilation or positive pressure ventilation. The former includes negative pressure body ventilators like the iron lung, cuirass, pneumosuit, pneumowrap, pancho, and other...
tank designs. These can provide sufficient assisted tidal volumes for effective alveolar ventilation, but when they are used during sleep like EPP/DP, they tend to cause obstructive sleep apneas that can necessitate concomitant continuous positive airway pressure (CPAP). They are impractical for daytime use. It is often more effective and comfortable to switch from nocturnal body ventilator or EPP/DP use to NIV. Negative pressure body ventilators are also rarely if ever as effective as NIV. Over 40 body ventilator and EPP/DP patients with no ventilator-free breathing ability (VFBA) have been transitioned to NIV to be freed from or remain free of tracheostomy.

The inspiratory muscles can also be aided/supported by applying positive pressure to the airways during or for inspiration. This can be done by intermittent positive pressure ventilation or by bi-level positive airway pressure (PAP). Both can be used noninvasively (NIV) or via invasive tubes.

**POSITIVE PRESSURE VENTILATION MODES**

**Passive Circuit Ventilation.** CPAP. CPAP and bi-level PAP are delivered via passive ventilator circuits (i.e., circuits without exhalation valves). These modes provide continuous air flow to dissipate CO2 during exhalation. CPAP does not assist inspiratory muscle function. It acts as a pneumatic splint to maintain airway patency and to increase functional residual capacity. It is rarely, if ever, indicated for patients with neuromuscular weakness.

**Bi-level PAP.** Bi-level PAP provides ventilatory assistance as a function of the inspiratory (I)PAP vs. expiratory (E)PAP difference (span). That is, an IPAP/EPPAP of 20/6 cm H2O assists inspiration almost as much as 14/0 cm H2O, but 14/0 cm H2O is more comfortable and increases intrathoracic pressures less. EPPAP of 0 cm, though cannot be provided by bi-level devices (“BiPAP” units). Bi-level PAP devices can provide full ventilatory support for NMD/SCI patients with normal lung compliance when it is used at IPAP/EPPAP spans of ≥18 cm H2O. Because they are necessarily pressure cycled, air stacking for lung volume recruitment is not possible when they are used. This prevents the lung volumes needed to increase voice volume and cough, diminish atelectasis, and maintain pulmonary compliance. Most devices also lack internal batteries, and for all these reasons they are impractical for daytime use, especially during meals.

Low-span (inspiratory PAP minus expiratory PAP spans <10 cm H2O) bi-level PAP becomes inadequate for ventilator support as inspiratory muscles weaken. Bi-level PAP, in general, is impractical for use via a mouthpiece, because it requires alternating current and its necessarily continuous air flow is uncomfortable.

**Proportion Assist Ventilation.** Proportion assist ventilation (PAV) supplements patients’ spontaneous tidal volumes as an inverse function of patient effort. It is a variant of bi-level PAP with all its inadequacies. Because NIV is an “open system,” NIV users can take any amount of the volume or pressure being delivered, and they naturally adjust their own supplemented volumes as needed. Because supplemental O2 and sedative medications must be avoided during sleep, the patient’s ventilation drive reflexively controls the amount of insufflation delivered to the lungs, rendering PAV unnecessary. As for any bi-level unit, PAV is also impractical for daytime use.

**Active Circuit Ventilation.** Synchronized intermittent mandatory ventilation (SIMV) and control ventilation are delivered via active circuits that have exhalation valves to dissipate CO2, so there is no need for airflow during exhalation. Thus, any holes in nasal interfaces used for NIV on active circuits generally need to be covered or plugged.

**Synchronized Intermittent Mandatory Ventilation (SIMV).** The mode SIMV can supplement patient breaths with pressure support and provide a back-up delivery of set volumes of air. Decreasing SIMV volumes and triggered breath pressure support levels along with supplemental O2 delivery and “permissive hypercapnia” is a common ventilator “weaning” strategy. Although this can be appropriate for managing lung/airways disease patients who have relatively poor prognoses, permitting hypercapnia is inappropriate for NMD/SCI patients, because it dulls ventilatory drive, renders NIV less effective during sleep, and ultimately exacerbates hypercapnia. Because most intubated and tracheostomized NMD/SCI patients can be extubated and decannulated without being “weaned” from ventilatory support, this mode is unnecessary.

**Volume Control Ventilation.** With volume control ventilation, the volumes delivered and the back-up rate are set. For older children and adults, volumes delivered typically range from 700 to 1500 ml along with physiologic back-up rates for age, typically 10–12/min. The high volumes permit the patient to take as much of the delivered volume as wanted for speech volume, coughing, assisting ventilation, and air stacking. Air stacking is lung volume recruitment by receiving and holding consecutively delivered volumes of air via manual resuscitator or volume-cycling ventilator with the glottis. The facilitation of air stacking for maintaining pulmonary compliance and maximizing cough flows and speech is the reason that assist-control
volume-cycling is the mode of choice for NIV for up to full, continuous ventilatory support.

**Pressure Control Ventilation.** Assist-control pressure mode with a physiologic back-up rate (rather than a high rate that promotes low-tidal-volume tachypnea) is the mode of choice when volume-cycling is ineffective because of excessive abdominal distension. For humans of all ages, lung volume/pressure characteristics are such that, for normally compliant lungs, delivered (tidal) volumes to pressures of about 18 cm H2O are required to maintain normal alveolar ventilation and physiologic breathing rates; that is, about 12 breaths of 500 ml each per minute for 6000 ml of minute ventilation for adults and normal PaCO2. Air stacking, however, is not possible with pressure cycling. Thus, when pressure-cycling needs to be used, lung volume recruitment needs to be provided by using a manual resuscitator or by high insufflation pressures from a mechanical insufflator–exsufflator (e.g., CoughAssist; Philips Respironics, Inc., Murrysville, Pennsylvania). Thus, although volume-cycling permits active lung volume recruitment for patients with glottis function, pressure-cycling permits passive lung recruitment when the glottis is too weak for effective air stacking.

**NONINVASIVE POSITIVE PRESSURE MECHANICAL VENTILATION**

Other than for CPAP, which does not assist ventilation, the modes of ventilation detailed earlier can be used for NIV. NIV for up to full, continuous ventilatory support is an alternative to tracheotomy and EPP/DP. It can be introduced in the hospital, clinic, or home setting. It is delivered via nasal and oral–nasal interfaces for nocturnal ventilatory support. These open systems require central nervous system reflexes to prevent excessive insufflation leakage during sleep. Supplemental oxygen and sedative medications can render NIV ineffective. Excessive insufflation leakage during sleep can be decreased by using a chin strap (Fig. 1), or better, by switching to closed nasal prong–lip-sealing systems (Liberty interface, Resmed, San Diego, California; and Hybrid interface, TeleflexMedicine, Research Triangle Park, North Carolina). Such interfaces deliver air via mouth and nose and require only minimal strap pressure.

NIV via a 15-mm angled mouthpiece is the most commonly used method for daytime ventilatory support. A mouthpiece can be kept between the teeth all day, but most patients have it fixed near the mouth by a flexible metal support arm attached to the wheelchair, or adjacent to sip and puff, chin, or tongue controls on a motorized wheelchair (Fig. 2). High delivered volumes permit the patient to vary the volume taken from each ventilator cycle to augment speech volume, cough, and air stack. Some neck movement and lip function are needed to grab the mouthpiece. The soft palate must move posterocranially to seal off the nasopharynx. In addition, the patient must open the vocal cords to maintain upper airway patency. These normal reflex movements may take time to learn for patients who have been using tracheotomy ventilation. Expertise in “noninvasive ventilation” requires experience in using NIV for

![FIGURE 1. An electrophrenic pacemaker (EPP) was implanted for this patient despite that fact that he was capable of daytime periods of ventilator-free breathing using accessory muscles. After implantation, he was placed on NIV, decannulated, and then permanently discontinued from EPP in favor of using mouthpiece NIV during the day and nasal NIV at night with a custom chin strap (seen here) to reduce insufflation leakage.](image1)

![FIGURE 2. Chin control motorized wheelchair with 15-mm angled mouthpiece adjacent to the mouth for easy access for continuous noninvasive ventilation (NIV).](image2)
around-the-clock support that includes daytime use of mouthpiece NIV.

**NIV Difficulties.** NMD/SCI patients with functional bulbar-innervated musculature but insufficient lip or neck strength to grab a mouthpiece invariably prefer to use nasal NIV diurnally via nasal prong systems rather than undergoing tracheostomy (Fig. 3). Other than for an uncontrollable seizure disorder or inability to cooperate, there are no contraindications to the long-term use of NIV. Severe bulbar dysfunction does not contraindicate NIV, but most ALS patients eventually require tracheostomy tubes for survival due to bulbar dysfunction. Abdominal distraction tends to occur sporadically in NIV users, and air is passed as flatus once the patient is mobilized in the morning. Aerophagia and abdominal distraction before introduction of NIV can be exacerbated and may result in it being ineffective. Placement of a gastrostomy tube to “burp” out the air can relieve the problem. Barotrauma is extremely uncommon in NMD/SCI NIV and MAC users.

**Intermittent Abdominal Pressure Ventilator.** Although negative pressure body ventilators are no longer desirable, the intermittent abdominal pressure ventilator (IAPV), or Exsufflation Belt (Respironics International, Inc., Murrysville, Pennsylvania), is a body ventilator that continues to be useful for daytime support. It involves intermittent inflation of an elastic air sac that is contained in a corset or belt worn beneath the patient’s outer clothing (Fig. 4). The sac is cyclically inflated by a positive pressure ventilator. Bladder inflation moves the diaphragm upward for a forced exsufflation. During bladder deflation, gravity returns the diaphragm for passive inspiration. A trunk angle of $\geq 30^\circ$ from the horizontal is necessary. If the patient has any inspiratory capacity or is capable of glossopharyngeal breathing (GPB), volumes of air can be autonomously added to each IAPV cycle. The IAPV augments tidal volumes by 300 ml to levels as high as 1200 ml; patients with $<1$ hour of VFBA often prefer it to NIV during daytime hours.

**EXPIRATORY MUSCLE AIDS**

Negative pressure applied directly to the airways during expiration as well as positive pressure applied to the thorax/abdomen (abdominal thrust) assists expiration and can increase cough flows. Electrical stimulation of abdominal muscles is not as effective as abdominal thrusts to increase cough flows for SCI patients.

Mechanically assisted coughing (MAC) is the combination of mechanical insufflation–exsufflation with an exsufflation-timed abdominal thrust. Insufflations to pressures of 40–60 cm H$_2$O, followed immediately by exsufflations to $-40$ to $-60$ cm H$_2$O, are most effective and are preferred. MAC can be provided via an oral–nasal mask, a simple mouthpiece, or a translaryngeal or tracheostomy tube, preferably with an inflated cuff. Whether via the upper airway or airway tube, routine deep suctioning misses the left main stem bronchus about 90% of the time, but MAC can clear both left and right airways without discomfort or trauma and is invariably preferable to invasive suctioning.

The goal is rapid, clinically full chest expansion followed by rapid lung emptying. One treatment consists of about 5 cycles of MAC followed by a brief period to clear the exsufflated secretions. Treatment continues until no further secretions are expelled, and associated oxyhemoglobin desaturation is reversed. Use can be required as often as every 30 minutes and almost around the clock during chest infections and for 1–2 days after extubation or decannulation.
Patient Evaluation. Pulmonary function testing (PFT) is not designed for the NMD/SCI patient. Oximetry, end-tidal carbon dioxide (EtCO₂) or transcutaneous carbon dioxide (TcCO₂) monitoring, spirometry for vital capacity (VC) measurements in sitting and supine positions and for measuring air stacking capacity (maximum insufflation efforts in sitting and supine positions and for measuring, spirometry for vital capacity (VC) measurements in sitting and supine positions and for measuring air stacking capacity (maximum insufflation capacity, maximum insufflation capacity, or MIC), and cough peak flow (CPF) analyses, both spontaneous and assisted by air stacking and abdominal thrust, are indicated. Unlike VC and nasal inflation pressure (NIF), which can be 0 despite intact bulbar function, assisted minus unassisted CPF and MIC minus VC are quantifiable and reproducible measures of glottis integrity and, therefore, bulbar function. SpO₂ <95% almost invariably indicates hypercapnia and airway congestion, and if the congestion is not expeditiously cleared, intrinsic lung disease, such as atelectasis or infiltrate can develop.

EtCO₂ and SpO₂ monitoring can be performed during sleep if the patient’s symptoms are questionable and the clinician seeks additional data to convince the patient to try nocturnal NIV. EtCO₂ or TcCO₂ and SpO₂ sleep monitoring are less expensive and more appropriate than polysomnography performed without EtCO₂ monitoring because the latter is not programmed to distinguish obstructive apneas from inspiratory muscle dysfunction. Although polysomnographic bi-level PAP titration is popular and may assure more restful sleep, because the VC will continue to decline, it must be repeated frequently. Less than full ventilator support settings are inadequate for optimal nocturnal inspiratory muscle rest or full ventilatory support when needed.

Critical Care. Most acute SCI patients who develop ventilatory failure do so during the first week of hospitalization. For uncomplicated patients, ventilatory failure and intubation can be avoided with noninvasive management (Fig. 5). Such SCI and MND patients’ VCs are measured every 8 hours. If they are decreasing, a portable ventilator is made available for NIV at full support settings to relieve dyspnea and avert acute ventilatory failure. Because no patient can develop ARF with SpO₂ ≥95% in ambient air, any SpO₂ <95% is reversed by NIV and/or MAC. No supplemental O₂ is administered unless desaturation cannot be reversed, indicating acute lung disease, for which the patient will likely need to be intubated.

Patients with NMD or SCI can nevertheless develop pneumonia and ARF and require intubation. However, even when patients are unweanable, extubation can almost invariably be accomplished for all but advanced bulbar ALS patients. Airway complications are more frequent for intubated patients who undergo tracheotomy than when simply maintaining intubation for up to 30 days. Medically uncomplicated but unweanable patients can remain intubated for ≥1 month to facilitate surgical procedures and to clear lung pathology to satisfy criteria for extubation to NIV and MAC (Table 1). Many unweanable patients have been intubated for over 1 month, even for 77 days, before being transferred for successful extubation to NIV. Avoiding tracheostomy ventilation decreases the risk of lifetime institutionalization. Unfortunately, most acute SCI patients’ conditions are complicated by traumatic brain injury, severe chest trauma, orthopedic and medical conditions, severe anxiety, and/or long-term narcotic therapy for pain management that dulls respiratory drive and can render NIV ineffective or necessitate intubation for >1 month.

Outcomes of Acute Management. We have now extubated 235 “unweanable” patients with inspiratory muscle dysfunction, 80 since our 2010 publication, without resorting to tracheotomy. The patients had typically failed extubation at other facilities that were not using full-setting NIV or

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<th>Table 1. Extubation criteria for unweanable patients.</th>
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<td>• Fully alert and cooperative, receiving no sedative medications.</td>
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<td>• Afebrile and normal white blood cell count.</td>
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<td>• PaCO₂ ≤40 mm Hg at peak inspiratory pressures &lt;30 cm H₂O on full ventilatory support and normal breathing rate, as needed.</td>
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<td>• Oxyhemoglobin saturation (SpO₂) ≥95% for ≥12 hours in ambient air.</td>
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<td>• All oxyhemoglobin desaturations &lt;95% reversed by mechanically assisted coughing and suctioning via translaryngeal tube.</td>
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<td>• Chest radiograph abnormalities cleared or clearing.</td>
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<td>• Air leakage via upper airway sufficient for vocalization upon cuff deflation.</td>
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MAC. Upon admission, supplemental oxygen was discontinued, and ambient air SpO₂ was often <95%. Ventilator settings were adjusted to normalize EtCO₂ and MAC applied via the translaryngeal or tracheostomy tube up to every 30 minutes to clear secretions and normalize SpO₂. The patients were extubated when specific criteria were met (Table 1). Patients who were unable to take food safely by mouth post-extubation underwent radiographically inserted or open gastrostomy without re-intubation. Patients with pre-extubation VCs of ≥250 ml regained ventilator free breathing ability (VFBA) by up to 3 weeks post-extubation, but most continued to require NIV during sleep. Post-extubation, patients weaned themselves when possible by taking fewer and fewer intermittent positive pressure ventilations as tolerated. Patients whose VCs remained <250 ml tended to require continuous NIV indefinitely.

Long-Term Management of Progressive Respiratory Dysfunction. Patients whose assisted CPF cannot exceed 5 L/s are prescribed oximeters and rapid access to MAC to maintain SpO₂ ≥95% during intercurrent infections to avoid pneumonia and ARF. Patients with intact bulbar musculature whose assisted CPF exceeds 5 L/s rarely require MAC.

Symptomatic patients with diminished VC are offered a trial of NIV to ease symptoms. When symptoms are questionable, home nocturnal EtCO₂ and SpO₂ monitoring showing CO₂ >50 cm H₂O and multiple hourly desaturations <95% indicates the need for a trial of nocturnal NIV. In general, patients with hypercapnia without concomitant SpO₂ <95% are usually asymptomatic.

When nocturnal-only NIV is no longer adequate and patients become dyspneic when attempting to discontinue its use in the morning, they continue nasal NIV into daytime hours and are switched to 15-mm angled mouthpiece interfaces. A finger oximeter can be worn and an SpO₂ alarm is set to 94% for severely hypercapnic patients. The goal is to maintain normal SpO₂ and, therefore, avoid symptomatic hypoventilation by autonomous breathing supplemented by NIV as needed. With time, the patient often becomes NIV-dependent 24 hours/day. We have managed 2000 patients using full-setting continuous NIV for up to 58 years with no VFBA and without resorting to polysomnography.

Ishikawa and colleagues reported long-term outcomes of noninvasive vs. tracheostomy management of Duchenne muscular dystrophy (DMD). For 21 consecutively managed patients undergoing tracheostomy from 1984 to 1991 mean survival was to 28.1 ± 8.3 years of age, with 3 still alive for a Kaplan–Meier life expectancy of 30.7 years. For 88 patients consecutively managed by NIV and MAC from 1991 to the 2010, however, life expectancy was 39.6 years (P < 0.001). The 88 NIV users began nocturnal use with a VC of 673 ± 320 ml at 18.9 ± 3.3 years of age and used it for 5.4 ± 3.3 years before 56 required full-time NIV once their VCs became 328 ± 194 ml at 24.7 ± 4.6 years of age.

In 2006, Toussaint et al. reported noninvasive management of 42 DMD patients aged 15–33 years who required nocturnal nasal NIV and daytime mouthpiece NIV. Survival rates were 88%, 77%, 58%, and 51% after 1, 3, 5, and 7 years, respectively, with a mean survival of 31 years. Kohler et al. reported a mean survival expectation of 35 years for noninvasively managed DMD patients. Eighteen patients became continuously NIV-dependent, including 1 for 10.1 years. Bach and Martinez described 101 DMD patients who were continuously NIV-dependent for a mean of 7.4 ± 6.1 years to age 30.1 ± 6.1 years, with 56 still alive. Twenty-six of the 101 became continuously dependent without hospitalization. Eight tracheostomized users were decannulated to NIV. Thirty-one consecutive unweanable intubated DMD patients were extubated to NIV.

Unlike the Ishikawa study in which no patient underwent tracheostomy, because the primary investigators extubated every acutely ill patient without it, Soudon et al. reported 3 patients, Bloch et al. reported 7 patients, and Bach et al. reported 4 patients who eventually underwent tracheostomy when they were intubated for acute illness and no longer had access to clinicians adept at NIV and MAC. Sixteen other centers reported 284 continuously NIV-dependent patients with DMD. Although other publications reported a brief statistical prolongation of survival by nocturnal-only use of low-span bi-level PAP in DMD, no other publications have reported survival by continuous NIV dependence.

Considering ALS, 335 patients from 11 centers became continuously NIV-dependent for means of 2–11 months (up to 8 years) before bulbar muscles deteriorated to cause continuous saliva aspiration and decreased baseline SpO₂, and necessitated tracheostomy for survival. This amounted to 25% to 42% of ALS patients requiring NIV continuously without hospitalization. With regard to ALS, although there have been numerous reports of brief increases in tracheostomy-free ALS survival by using nocturnal low-span bi-level PAP, the mechanism for this may be in the slightly deeper breaths increasing cough flows to expulse airway secretions. Prolonged ALS survival by continuous NIV support has only been described in 2 studies. In
one study, Bach et al. also reported the extubation of 11 ALS patients who could not pass spontaneous breathing trials either before or after extubation.

Considering typical SMA type 1, for which tracheostomy-free life expectancy is <18 months, 34 patients with no VFBA were reported who depended on continuous NIV, including 15 >10 years of age with no VFBA since as early as 4 months of age. Seventy-two SMA type 1 NIV users had a mean age of 86.1 (range 13–196) months, and 13 died at a mean age of 52.3 (range 13–111) months. Continuous NIV support rather than tracheostomy has also been reported for 18 other SMA type 1 patients at 5 other centers.

Aging SCI patients can also develop the need for nocturnal NIV and even continuous NIV support. As for NMD patients, when CPF is poor, they too are very susceptible to episodes of pneumonia triggered by intermittent chest colds that, if not managed by MAC, can result in ARF.

Glossopharyngeal Breathing. The use of the glottis to piston air into the lungs, can provide normal alveolar ventilation for individuals with no VC or VFBA when the ventilator fails during sleep. GPB and normal PaCO₂ throughout daytime hours by glossopharyngeal breathing and maximum glossopharyngeal single breath capacity of 3500 ml.

The glottis closes with every glossopharyngeal “gulp.” One breath usually consists of 6–9 gulps of 40–200 ml each (Fig. 6). During the training period, the efficiency of GPB can be monitored by spirometrically measuring the milliliters of air per gulp, gulps per breath, and breaths per minute. A training manual and numerous videos are available, the best of which for SCI patients was produced in 1999. Although non-bulbar ALS patients are able to do it, it has not yet been shown to be useful for this population.

Because GPB is rarely useful in the presence of an indwelling tracheostomy tube and is never useful when a patient is being ventilated through one, the safety and versatility it affords are additional reasons to eliminate tracheostomy in favor of noninvasive management.

Decannulation of Unweanable Patients. The ability to generate ≥2.7 L/s of assisted CPF, an important determinant for adequate bulbar function and airway patency, indicates a potential for successful decannulation and effective use of MAC irrespective of VFBA. An MIC/VC difference of >0 ml also indicates glottis closure and offers good potential for decannulation. Over 50 unweanable SCI patients were successfully decannulated before 1991. These patients subsequently used continuous NIV for up to 38 years without re-tracheostomy or even re-hospitalization for many cases.

DMD, non-bulbar ALS, and patients with other NMDs have also been decannulated despite requiring continuous ventilatory support. The principles of decannulating unweanable patients to NIV are essentially the same as those for extubation, except they require a pressure ostomy dressing during continuous NIV to prolong survival by >3000 patient-years without tracheostomies. Four of the centers reported having routinely decannulated >200 unweanable patients with functioning bulbar musculature.

Phrenic and Diaphragm Pacing. In 2009, laparoscopic implantation of electrodes for diaphragm pacing (DP) was reported, where electrodes were implanted directly into the motor points of the diaphragm for 50 SCI and 38 ALS patients. In October 2011, the FDA approved its “compassionate use” for ALS. However, although it can be effective for high-level SCI patients, it has been ineffective for all 8 ALS patients we have monitored.

In 1997, guidelines were published for the management of high-level, ventilator-dependent

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<th>Level of Lesion</th>
<th>VC (ml)</th>
<th>Function</th>
<th>Ventilation</th>
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<tr>
<td>Above C1</td>
<td>&lt;200</td>
<td>–</td>
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<tr>
<td>C1–C2</td>
<td>&lt;200</td>
<td>+</td>
<td>TIV, TIV</td>
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<tr>
<td>C3 or lower</td>
<td>&gt;200</td>
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<td>NIV/AAPV, NIV</td>
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VC, vital capacity; TIV, intermittent positive pressure ventilation via tracheostomy; EPP, electrophrenic pacing; DP, diaphragm pacing; NIV, noninvasive intermittent positive pressure ventilation; APV, intermittent abdominal pressure ventilator.
SCI patients who have little or no VFBA (Table 2) \(^{42}\): Category 1—Patients with lesions extending into the brainstem who have severe bulbar-innervated muscle impairment and inability to protect the airways require tracheostomy tubes for airway protection and may benefit from EPP or DP for ventilatory support. Category 2—High-level patients with adequate airway protection but without the ability to grab a mouthpiece for NIV can use MAC to clear the airways and EPP or DP along with nasal NIV around-the-clock and be safely decannulated. Category 3—Patients with or without VFBA who can grab a mouthpiece can be decannulated to continuous NIV.

Many Category 3 patients have been successfully decannulated to continuous NIV. \(^{12,27,43}\) and, recently, the value of EPP was reported to permit decannulation of ventilator-dependent SCI patients as well. \(^{46}\) Thus, while Category 1 and 2 SCI patients with no VFBA and VC <250 ml can benefit from EPP or DP, neither is necessary for the decannulation of Category 3 patients. \(^{11,12}\) Long-term complications of EPP/DP can include phrenic compression neuropathies (for EPP), myopathic changes of the diaphragm, \(^{44}\) and infection. \(^{27,45}\) Also, because EPP/DP patients are generally left with tracheostomy tubes, they suffer complications of the latter as well. \(^{46}\)

**Diaphragm Pacing and ALS.** A 1999 consensus group of the National Association for Medical Direction of Respiratory Care cited a VC of 50% as an indication for nocturnal NIV. \(^{47}\) This figure is also being used for DP placement for ALS patients, \(^{27}\) despite absence of symptoms or any benefit on survival or retention of pulmonary function. Indeed, the VC of ALS patients have been reported to taper with or without the use of NIV or DP. \(^{38,49}\) Of our 8 DP patients, 6 died at 4–17 months after DP placement, 1 underwent tracheostomy for ventilatory support after 17 months, and rates of decrease in VC post–DP placement ranged from 3.7% to 20% per month.

The chief difference between EPP/DP use in SCI vs. ALS is that, for high-level SCI patients whose VC's are inadequate to sustain alveolar ventilation and who breathe at or close to inspiratory capacity, EPP/DP provides volumes that can greatly exceed inspiratory capacity; that is, tidal volumes that can normalize alveolar ventilation. Thus, SCI patients with no VFBA can turn off the ventilator and use EPP/DP alone. For ALS patients with VC's >2 liters (about 50% of predicted normal), DP does not deliver >25% of that which can be attained by the patient’s own inspiratory muscles. Further, because the patient’s breathing is not synchronized with the DP, the patient will potentially breathe as much with it as against it.

**CONCLUSIONS**

Noninvasive respiratory management, including NIV and MAC, can be used to avoid respiratory failure, hospitalizations, and tracheotomy for patients with NMDs and SCI and to permit extubation and decannulation of unweanable patients, provided that bulbar muscle function is adequate to protect the airways. All patients who have used both invasive and noninvasive ventilation for ventilatory support prefer the latter. \(^{50}\) EPP/DP is only indicated for high-level SCI patients who have no ventilator-free breathing ability. Such patients can also be offered decannulation to avoid the long-term complications and discomfort of tracheostomy. The NMD and SCI clinician needs to understand the noninvasive approach and its potential outcomes to be able to employ consultants who can achieve them.

This monograph was edited and approved by committees of the AANEM. It did not undergo further peer review by Muscle & Nerve.

**REFERENCES**


