Review Article

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Noninvasive Respiratory Management for Patients with Spinal Cord Injury and Neuromuscular Disease

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The purpose of this article is to describe noninvasive respiratory management for patients with neuromuscular respiratory muscle dysfunction (NMD) and spinal cord injury (SCI) and the role of electrophrenic pacing (EPP) and diaphragm pacing (DP) in this respect. Long term outcomes will be reviewed and the use of noninvasive intermittent positive pressure ventilation (NIV), MAC, and EPP/DP to prevent pneumonia and acute respiratory failure, to facilitate extubation, and to avoid tracheotomy will be evaluated. Although ventilator dependent patients with most NMDs and high level SCI can be indefinitely managed noninvasively, most ALS patients can be managed for a limited time by continuous NIV before tracheostomy is necessary for survival.
Glossopharyngeal breathing (GPB) can be learned by patients without any autonomous breathing ability and used by them in the event of ventilator/EPP/DP failure or loss of interface access. EPP/DP can maintain alveolar ventilation for high level SCI patients when they cannot grab a mouth piece to use NIV.

Key words: Glossopharyngeal breathing, Assisted cough, Mechanical insufflation-exsufflation, Spinal cord injury, Tetraplegia, Respiratory therapy, Noninvasive mechanical ventilation, Electrophrenic pacing, Diaphragm pacing

Abbreviations: CPAP: *continuous positive airway pressure ,* ***DP*** *: diaphragm pacing by implantable electrodes implanted into diaphragm motor points,* ***EtCO2:*** *end-tidal CO2 mm Hg,* ***EPAP:*** *expiratory positive airway pressure,* ***EPP:*** *electrophrenic pacing of diaphragm by electrodes placed on phrenic nerves,* ***GBP:*** *glossopharyngeal breathing ("frog breathing") – the use of the glottis to piston boluses of air into the lungs,* ***IAPV:*** *intermittent abdominal pressure ventilator,* ***MAC:*** *mechanically assisted coughing, the combination of mechanical insufflation-exsufflation with an exsufflation-timed abdominal thrust,* ***NIV****: noninvasive intermittent positive pressure ventilation,* ***NMD:*** *neuromuscular respiratory muscle dysfunction,* ***PAP:*** *positive airway pressure,* ***SpO2:*** *oxyhemoglobin saturation by pulse oximeter,* ***VC:*** *vital capacity*

Inspiratory and Expiratory Muscle Aids

EPP/DP and negative pressure body ventilators can both assist with effective alveolar ventilation but cause obstructive sleep apneas, necessitating concomitant continuous positive airway pressure (CPAP). However, negative pressure body ventilators are rarely as effective as NIV. NIV also assists inspiratory muscles and can be used comfortably during sleep as well as during the day. The only body ventilator that will be considered here is the intermittent abdominal pressure ventilation (IAPV).

Negative pressure applied directly to the airways during expiration as well as positive pressure applied to the thorax / abdomen (abdominal thrust) assists expiration. Both deep positive pressure insufflations and a mechanical exsufflation and abdominal thrust are needed to increase cough flows.

Noninvasive Intermittent Positive Pressure Ventilation (NIV)

NIV for continuous ventilatory support is an alternative to tracheotomy and EPP / DP (1). It is delivered via a lip sealing mouth piece, nasal, and oral-nasal interfaces for nocturnal ventilatory support. Supplemental oxygen and sedative medications can render NIV ineffective.

NIV via mouth piece is the most important method of daytime ventilatory support (Figure 1). A mouth piece can be kept between the teeth all day,(2) but most patients have it fixed near the mouth by a flexible metal support arm attached to the wheelchair (3,4). Some neck movement and lip function are needed, and the patient must open the vocal cords to maintain upper airway patency.

Figure 1. Continuously ventilator dependent 45-year-old woman with non-bulbar amyotrophic lateral sclerosis who was decannulated to continuous mouth piece ventilation during daytime hours and nasal ventilation for sleep.

NIV Difficulties

The NMD / SCI patients with bulbar functioning whose lips are too weak to grab a mouth piece for daytime NIV prefer to use nasal NIV diurnally rather than undergo tracheostomy (Figure 2).(1)

Figure 2. 35-year-old lawyer with Duchenne muscular dystrophy decannulated after 6 months of continuous ventilatory support via tracheostomy with no measurable VC but with lips too weak to use mouth piece noninvasive ventilation. He, therefore, uses a Nasal-Aire Interface™ (InnoMed Technologies, Coconut Beach, Florida) during daytime hours that permits him to wear glasses.

Other than for an uncontrollable seizure disorder or inability to cooperate, there are no contraindications to the long-term use of NIV or extubation to noninvasive ventilatory support (5). Barotrauma is extremely rare in NMD / SCI NIV and MAC users (6).

Ventilator Modes

CPAP

CPAP is not a ventilatory mode but acts as a pneumatic splint. It does not directly assist respiratory muscle function.

Bi-Level PAP

Bi-level positive airway pressure (PAP) can provide full ventilatory support for NMD / SCI patients with normal lung compliance when used at an inspiratory PAP, expiratory PAP spans of 18 cm H_2O or greater. Since they are necessarily pressure cycled, patients cannot air stack when using bi-level PAP.

Volume and pressure control ventilation

With volume control ventilation volumes are delivered with a set back-up rate. High delivered volumes of 700 –

1500 ml permit the adult patient to air stack, which improves pulmonary compliance and cough.

Body Ventilators

The IAPV or "Exsufflation Belt™" is a body ventilator that continues to be useful for daytime support (Figure 3). An elastic air sac is cyclically inflated, moving the diaphragm upward for a forced exsufflation. During bladder deflation, inspiration occurs passively. If the patient is capable of GPB, volumes of air can be autonomously added to each IAPV cycle.

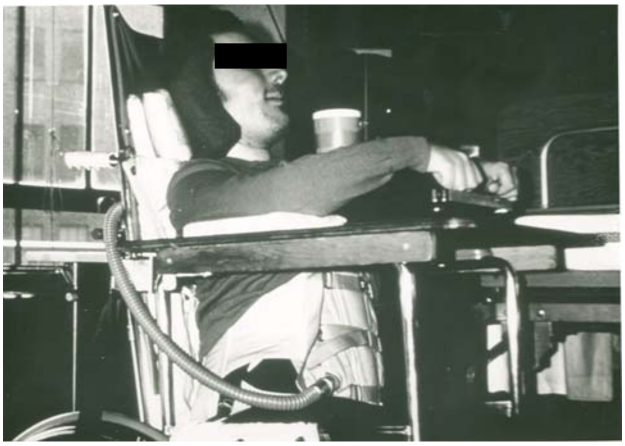

Figure 3. A 44-year-old with Duchenne muscular dystrophy using an intermittent abdominal pressure ventilator (IAPV) for daytime support and nocturnal noninvasive ventilatory support via a lip sealing mouth piece seen here with the hose of the portable ventilator entering his IAPV under his clothing.

Mechanically Assisted Coughing (MAC)

MAC is useful for resolving secretion retention. MAC can be provided via an oral-nasal mask, a simple mouthpiece, or via a translaryngeal or tracheostomy tube. Routine deep suctioning misses the left main stem bronchus about 90% of the time (7), but MAC flows can be effective in both right and left airways without discomfort (8).

A mechanical in-exsufflator can be manually or automatically cycled. Manual cycling to synchronize with the patient's breathing is necessary for patients who cannot cooperate with automatic cycling. Use can be required as frequently as every 30 minutes almost around the clock during infection.

Evaluation and Management in the Acute Care Setting

For the NMD/SCI patient, oximetry, end-tidal CO_2 ($EtCO_2$) or transcutaneous carbon dioxide monitoring, spirometry, and cough peak flow analyses are indicated for this population. SpO_2<95% almost invariably indicates hypercapnia and airway congestion, and this may result in lung infection or atelectasis if not cleared by mechanically assisted coughing (MAC). Thus, patients should be treated by NIV and/or MAC without supplemental O_2 to maintain oxyhemoglobin saturation (SpO_2) greater than 94%. If the SpO_2 remains below 95% despite NIV and MAC, the patient likely has pneumonia and intubation may soon be required.

Many unweanable patients have been intubated for over 1 month, even for up to 77 days, before being transferred for successful extubation to NIV (5). One month of intubation with excellent potential to be extubated to NIV and MAC (given bulbar function), is preferable to tracheotomy, and possibly a lifetime of institutionalization.

Outcomes of Acute Management

Over 215 "unweanable" patients with NMD/SCI were extubated without resort to tracheotomy (9,11). Upon admission, supplemental oxygen was invariably discontinued, ventilator settings were adjusted to normalize $EtCO_2$, with MAC up to every 30 minutes to clear secretions and normalize SpO_2. When other extubation criteria were also satisfied (Table 1), the patients were successfully extubated. Extubation success was defined as not requiring re-intubation during the hospitalization (9,11).

Table 1.

Extubation Criteria for Unweanable SCI Patients
• Fully alert and cooperative, receiving no sedative medications
• Afebrile and normal white blood cell count
• $PaCO_2$ 40 mm Hg or less at peak inspiratory pressures less than 30 cm H2O on full ventilatory support and normal breathing rate, as needed
• Oxyhemoglobin saturation (SpO2) ≥ 95% for 12 hours or more in ambient air
• All oxyhemoglobin desaturations below 95% reversed by mechanically assisted coughing and suctioning via translaryngeal tube
• Chest radiograph abnormalities cleared or clearing
• Air leakage via upper airway sufficient for vocalization upon cuff deflation

Long-Term Management of Slowly Progressive Respiratory Dysfunction

When multiple hourly desaturations below 95% occur, and home $EtCO_2$ measures over 50 cm H_2O, a trial of nocturnal NIV may help clear symptoms and normalize blood gases. In general, patients with hypercapnia without concomitant SpO_2<95% are usually asymptomatic or have questionable symptoms that are cleared by nocturnal NIV.

Daytime $EtCO_2$ measurement along with SpO_2 sleep monitoring is more appropriate than polysomnograms because the latter do not evaluate inspiratory muscle dysfunction. Thus, $EtCO_2$, SpO_2 measurement, cough peak flows, and spirometry for sitting and supine VC and air stacking capacity measurements (12), GPB maximum volumes, and air stacking to capacity should be performed at all patient visits. With aging and continued loss of VC, patients often develop the need for around-the-clock noninvasive ventilatory support (13).

Lung and Chest Wall Mobilization

Regular mobilization is required to prevent chest-wall contractures and lung restriction. This can only be achieved by active (air stacking) or passive deep insufflations (12). Despite daily air stacking to up to 70 cm H2O pressures for over 1500 patients, only two cases of pneumothorax have been reported in association with ventilator use (5).

Patients who cannot close the glottis and, therefore, cannot air stack, can be passively insufflated to pressures of 40 to 70 cm H_2O by manual resuscitator with the exhalation valve blocked. The primary objectives of lung volume recruitment therapy are to maintain or improve pulmonary compliance and cough, and to diminish atelectasis.

Glossopharyngeal Breathing

Patients with intact bulbar function can use GPB to go off the ventilator or when the ventilator fails day or night. Two-thirds of SCI patients with no autonomous breathing ability can use GPB for autonomous breathing (3,4). The technique involves the use of the glottis to add to an inspiratory effort by pistoning boluses of air into the lungs. It can be equally useful for NMD patients except for those with bulbar-ALS (3,4,14).

Decannulation of Unweanable Patients

The principles of decannulating unweanable patients are essentially the same as those for extubation except that they require a pressure ostomy dressing until ostomy closure to use noninvasive ventilatory support.

Phrenic (EPP) and Diaphragm Pacing (DP)

Electric stimulation of the phrenic nerves (EPP) was demonstrated to support ventilation in 1948 (15). A total of 700 pacemakers were implanted, yet relatively few were implanted for SCI patients with intact phrenic nerves by comparison to COPD patients and others for whom it was useless. This is similar to the current situation for DP that is futilely being used for ALS.

The chief difference between EPP/DP use in SCI vs. ALS is that for high level SCI patients EPP/DP provides volumes that can greatly exceed inspiratory capacity. SCI patients with no autonomous breathing ability can turn off the ventilator and use EPP/DP instead. For ALS, most patients do not have enough inspiratory capacity and cannot turn off the ventilator.

CONCLUSION

Noninvasive respiratory management including NIV and MAC can be used to avoid respiratory failure, hospitalizations, and tracheotomy for patients with NMDs and SCI who have functioning bulbar musculature and can be used to extubate and decannulate patients. EPP/DP is only indicated at the present time for high level SCI patients who have no autonomous breathing ability. Thus, a paradigm shift is required for optimal rather than invasive management and lifetime institutionalization of SCI and NMD patients who require ventilatory support. To accomplish this, the physician must understand the noninvasive approach.

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