Respiratory Muscle Aids to Avert Respiratory Failure and Tracheostomy

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Abstract

The purpose of this article is to describe the use of noninvasive inspiratory and expiratory muscle aids to prevent ventilatory insufficiency and failure, and to permit the extubation and tracheostomy tube decanulation of "unweanable" patients. Noninvasive airway pressure aids can provide up to continuous ventilator support for patients with little or no vital capacity and can provide for effective cough flows for patients with severely dysfunctional expiratory muscles. An April 2010 consensus of clinicians from 22 centers in 18 countries reported 1623 spinal muscular atrophy type 1 (SMA1), Duchenne muscular dystrophy (DMD), and amyotrophic lateral sclerosis (ALS) NIV users, of which 760 developed continuous ventilator dependence to prolong survival by over 3000 patient-years without tracheostomies. Four of the centers routinely extubated unweanable DMD patients so that none of their over 250 such patients has undergone tracheotomy. This approach is now making inroads into Canada via centers in Ottawa and Montreal.

What Are Physical Medicine Respiratory Muscle Aids?

Inspiratory and expiratory muscle aids are devices and techniques that involve the manual or mechanical application of forces to the body, or intermittent pressure changes to the airway, to assist inspiratory or expiratory muscle function. The devices that act on the body include body ventilators that create pressure changes around the thorax and abdomen. Negative pressure applied to the airway during expiration assists coughing, just as positive pressure applied to the airway during inhalation (NIV) assists the inspiratory muscles. Continuous positive airway pressure (CPAP) does not assist ventilation and is not useful for patients with primarily ventilatory impairment.

Patient Evaluation

Patients with diminished ventilatory reserve who are able to walk commonly complain of exertional dyspnea. Eventually, morning headaches, fatigue, sleep disturbances, and hypersomnolence develop (2). For wheelchair users, symptoms may be minimal except during intercurrent respiratory infections when they complain of anxiety, inability to fall asleep, and dyspnea. Patients are observed for tachypnea, paradoxical breathing, hypophonia, nasal flaring, use of accessory respiratory musculature, cyanosis, flushing or pallor, and airway secretion congestion. Lethargy and confusion signal CO_2 narcosis.

Evaluation necessitates 4 items: a spirometer, peak flow meter, capnograph, and oximeter. The VC is measured in sitting and supine positions. The VC difference should be less than 7%. Since hypoventilation is worse during sleep, the supine rather than sitting position VC is the most important indicator of ventilatory dysfunction. When it is greater than 20% orthopnea often indicates the need for nocturnal NIV. Patients wearing

thoracolumbar bracing should have the VC measured both with the brace on and off, since a good fitting brace can increase VC - whereas a poorly fitting one can decrease it. Spirometry is also useful for monitoring progress with GPB and air stacking, that is, retention of a maximum lung volume of aid delivered by manual resuscitator or volume cycling ventilator that can be held by the glottis. The maximum volume is termed the maximum insufflation capacity (MIC). Patients who learn GPB can often air stack consecutive GPB gulps to or beyond the MIC (3). A nasal interface or lipseal can be used for air stacking when the lips are too weak for effective air stacking via the mouth.

Cough peak flows (CPF) are measured using a peak flow meter (Access Peak Flow Meter, Healthscan Products Inc., Cedar Grove, NJ). CPF of 160 L/m are the minimum needed to cough effectively (3), and this is the best indicator for tracheostomy tube removal irrespective of remaining pulmonary function. Indeed, 40% of patients with ALS can survive despite continuous ventilator dependence using strictly noninvasive aids (1). Patients with VCs less than 1,500 ml have assisted CPF measured from a maximally stacked volume of air and with an abdominal thrust delivered simultaneously with glottic opening.(4) Coughing from a deep air stacked volume with a concomitantly applied abdominal thrust is termed a manually assisted cough.

For the stable patient without intrinsic pulmonary disease, arterial blood gas sampling is unnecessary. Besides the discomfort, 25% of patients hyperventilate as a result of anxiety or pain during the procedure. (5) Noninvasive continuous blood gas monitoring, including capnography and oximetry, yield more useful information, particularly during sleep.

While all clearly symptomatic patients with diminished lung volumes require a trial of NIV to ease symptoms, if symptoms are questionable, nocturnal noninvasive blood gas monitoring can be performed. The oximeter and the capnograph, which measures end-tidal pCO_2, must be capable of summarizing the data. (2) These studies are most conveniently performed in the home. Any questionably symptomatic patient with decreased VC, multiple nocturnal oxyhemoglobin desaturations below 95%, and elevated nocturnal $PaCO_2$ should also undergo a trial of nocturnal NIV. Since, in general, only patients improperly treated with supplemental O_2 develop CO_2 narcosis and ARF is generally caused by ineffective cough and airway secretion management, any patient finding that NIV use is more burdensome than symptoms of ventilatory insufficiency is told that it is alright to discontinue NIV and return for a re-evaluation in 3 to 6 months.

For symptomatic patients with normal VC, an unclear pattern of oxyhemoglobin desaturation, and no apparent carbon dioxide retention, sleep disordered breathing is suspected and a polysomnogram warranted.(6) Obesity-hypoventilation patients are treated with nocturnal ventilatory support, as are NMD patients, and not with CPAP. Polysomnography is not indicated for patients with decreased VC (NMD) because it is programmed to interpret every apnea and hypopnea as resulting from central or obstructive events rather than from inspiratory muscle weakness. Further, treatment of asymptomatic NMD patients on the basis of polysomnographic abnormalities neither prolongs life nor improves its quality.

The Intervention Objectives

The intervention goals are to maintain lung and chest-wall compliance and to promote normal lung and chest-wall growth for children, to maintain normal alveolar

ventilation around the clock, and to maximize CPF. The long-term goals are to avert episodes of ARF, especially during intercurrent chest infections; to avoid hospitalizations, and prolong survival without resorting to tracheotomy. Unweanable intubated and canulated patients can be extubated and decanulated to NIV and mechanically assisted coughing (MAC). All goals can be facilitated by evaluating, training, and equipping patients in the outpatient setting and at home.

Long-Term Management

Goal One: Maintain pulmonary compliance, lung growth, and chest-wall mobility

Pulmonary compliance is diminished because the patient can not expand the lungs to predicted inspiratory capacity. As the VC decreases, the largest breath one can take only expands a fraction of lung volume. Like limb articulations, regular mobilization is required to prevent chest-wall contractures and lung restriction. This can only be achieved by providing deep insufflations, air stacking, or nocturnal NIV (7). The extent to which the MIC exceeds VC (MIC-VC) objectively quantitated glottic, and therefore bulbar-innervated muscle integrity, and correlates with the capacity to use noninvasive aids rather than tracheostomy. (4) Patients who can not close the glottis and, therefore, can not air stack, must be passively insufflated using a CoughAssist™ (Respironics International Inc., Murrysville, Pa), pressure-cycling ventilator at pressures of 40 to 70 cm H_2O, or manual resuscitator with the exhalation valve blocked. The maximum passive insufflation volume can be termed the "Lung Insufflation Capacity" or LIC. (8)

The primary objectives of lung expansion therapy are to increase the VC and to maximize CPF, to maintain or improve pulmonary compliance, to diminish atelectasis, and to master NIV. In 282 spirometry evaluations of NMD patients for VC, MIC, and

LIC, the authors found mean values of 1131 ± 744 mL, 1712 ± 926 mL, and 2069 ± 867 mL, respectively. (8) The deeper lung volumes by air stacking also permitted patients to raise voice volume as desired.

Because any patient who can air stack is also able to use NIV, if such a patient is intubated for respiratory failure, he or she can more easily be extubated directly to continuous NIV regardless of ventilator-free breathing ability (VFBA). Extubation of patients without VFBA who are inexperienced in NIV can result in panic, ventilator dyssynchrony, asphyxia, and, at times, reintubation.

Before patients' VCs decrease to 70% of predicted normal, they are instructed to air stack 10 to 15 times, at least two or three times daily usually using a manual resuscitator. Because of the importance of air stacking, NIV is provided via ventilators using volume rather than pressure cycling, on assist/control mode.

Infants cannot air stack or cooperate with passive insufflation therapy. All babies with spinal muscular atrophy (SMA) type 1, infants with SMA type 2, and others with infantile NMD who have paradoxical chest-wall movement require nocturnal NIV to prevent pectus excavatum and promote lung growth as well as for ventilatory assistance. (9) In addition to nocturnal aid, deep insufflations may be possible by delivering air from a manual resuscitator via an oral-nasal interface and timing the air delivery to the child's breathing. Children can become cooperative with deep insufflation therapy by 14 to 30 months of age.

Goal Two: Maintain normal alveolar ventilation by inspiratory muscle assistance

Although the inspiratory muscles can be assisted by applying pressures to the body, negative pressure body ventilators cause obstructive apneas, are less effective than

NIV, and become increasing less effective with age and decreasing pulmonary compliance. (10) Blood gases improve dramatically when switching patients from them to NIV.

A body ventilator that continues to be useful is the intermittent abdominal pressure ventilator (IAPV) or "Exsufflation Belt™" (Respironics International Inc., Murrysville, PA). It involves the intermittent inflation of an elastic air sac that is contained in a corset or belt worn beneath the patient's outer clothing. The sac is cyclically inflated by a positive pressure ventilator. Bladder inflation moves the diaphragm upward to assist in expiration. During bladder deflation, gravity causes the abdominal contents and diaphragm to return to the resting position and inspiration occurs passively. A trunk angle of 30 degrees or more from the horizontal is necessary for it to be effective. If the patient has any inspiratory capacity or is capable of GPB, he or she can add volumes of air autonomously taken in to that taken in mechanically. The IAPV augments tidal volumes by 300 to as high as 1,200 mL; patients with less than one hour of breathing tolerance usually prefer it to using NIV during daytime hours.(11)

Noninvasive Intermittent Positive Pressure Ventilation (NIV)

NIV can be noninvasively delivered via lipseals, nasal, and oral-nasal interfaces for nocturnal ventilatory support. Mouthpiece and nasal IPPV are open systems that require the user to rely on central nervous system reflexes to prevent excessive insufflation leakage during sleep (2, 12), thus, supplemental oxygen and sedatives can render NIV ineffective. NIV should be introduced in the clinic or home setting.

There are numerous commercially available nasal interfaces (CPAP masks). Several should be tried and the patient should be encouraged to alternate their use.

Excessive insufflation leakage can be avoided by switching to the use of a closed noninvasive system such as using lipseal-nasal prong systems. Such interfaces deliver air via mouth and nose during sleep and require minimal strap pressure. This optimizes skin comfort and minimizes air (insufflation) leakage. Excessive leakage is also prevented by sustaining ventilatory drive by maintaining normal daytime CO_2 and avoiding supplemental O_2 and sedatives.

NIV via a 15 mm angled mouth piece is the most important method of daytime ventilatory support. Some patients keep the 15 mm angled mouthpiece between their teeth all day. (13) Most have the mouthpiece held near the mouth. A metal clamp attached to a wheelchair can be used for this purpose, or the mouthpiece can be fixed onto motorized wheelchair controls—most often, sip and puff, chin, or tongue controls. The ventilator is set for large tidal volumes, often 800 to 1500 mL. The patient grabs the mouthpiece with his mouth and supplements or substitutes for inadequate autonomous breath volumes. The patient varies the volume of air taken from ventilator cycle to ventilator cycle and breath to breath to vary speech volume and cough flows as well as to air stack to fully expand the lungs. Some neck movement and lip function are needed to grab the mouth piece and use it without leaking air. The soft palate must move in the posteriocranial direction to seal off the nasopharynx. In addition, the patient must open the glottis and vocal cords, dilate the hypopharynx, and maintain airway patency. These normally reflex movements may require a few minutes to relearn for patients who have been receiving ventilation via a tracheostomy tube. (14)

Nasal NIV is most practical for nocturnal use but it is also indicated for infants and for those who can not grab or retain a mouth piece because of oral muscle weakness,

inadequate jaw opening, or insufficient neck movement. Continuous nasal NIV is, nevertheless, a viable and desirable alternative to tracheostomy. (2) Nasal NIV users learn to close their mouths or seal off the oropharynx with their soft palates and tongues to prevent oral insufflation leakage.

Suboptimal humidification dries out and irritates nasal mucous membranes, causes sore throat, and results in vasodilatation and nasal congestion. Increased airflow resistance to 8 cm H_2O can be caused by the loss of humidity that is due to unidirectional airflow with expiration via the mouth during nasal CPAP or NIV. (15) This can be reduced by warming the inspired air to body temperature and humidifying it using a hot-water bath humidifier. (15) Decongestants can also relieve sinus irritation and nasal congestion. Switching to lip-seal-only interface can relieve most if not all difficulties associated with nasal NIV. Other than perhaps for an uncontrollable seizure disorder and inability to cooperate, there are no contraindications to the long-term use of noninvasive inspiratory muscle aids.

Abdominal distention tends to occur sporadically in NIV users. The air usually passes as flatus once the patient is mobilized in the morning. When severe, however, it can increase ventilator dependence and necessitate a rectal tube to decompress the colon or a nasogastric or gastrostomy tube to burp out the air.

Despite aggressive lung mobilization and expansion three times daily, often to over 60 cm H_2O pressures and along with NIV support for over 50 years in many cases, we have had one case of pneumothorax in over 1000 NIV users. Although often described as a complication or limiting factor for NIV, secretion encumbrance most often results from failure to use MAC.

GOAL THREE: ASSIST EXPIRATORY MUSCLES TO AUGMENT COUGH FLOWS

Manually assisted coughing is the use of air stacking for any patient with less than 1500 ml of VC to precede an abdominal thrust timed to glottic opening. With the higher lung volumes by air stacking, assisted CPF of 4.3 ± 1.7 L/sec were obtained by comparison with 2.5 ± 2.0 L/sec unassisted.(4) In 364 evaluations of our NMD patients able to air stack, the mean VC in the sitting position was 996.9 mL, the mean MIC was 1647.6 mL, and although CPFs were 2.3 L/s (less than 2.7 L/s or the minimum needed to eliminate airway secretions) mean assisted CPF were 3.9 L/s. This is the difference between coughing effectively to prevent pneumonia and ARF or not. (16) The inability to generate 160 L/m of assisted CPF despite having a VC or MIC greater than 1 L indicates upper-airway obstruction often due to severe bulbar-innervated muscle dysfunction and should be evaluated by laryngoscopy and reversible lesions corrected surgically.

Mechanically assisted coughing (MAC) is the combination of the use of mechanical insufflation-exsufflation (CoughAssist™) with an exsufflation-timed abdominal thrust. Deep insufflations followed immediately by deep exsufflations at pressures of 40 to –40 cm H_2O are usually the most effective and preferred. MAC can be provided via an oral-nasal mask, a simple mouthpiece, or via a translaryngeal or tracheostomy tube. When delivered via the latter, the cuff, when present, should be inflated. The CoughAssist™ can be manually or automatically cycled. Manual cycling facilitates caregiver-patient coordination of inspiration and expiration with insufflation

and exsufflation, but it requires hands to deliver an abdominal thrust, to hold the mask on the patient, and to cycle the machine.

One treatment consists of about five cycles of MAC followed by a short period of normal breathing or ventilator use to avoid hyperventilation. Insufflation and exsufflation times are adjusted to provide maximum chest expansion and rapid lung emptying. In general, 2 to 4 seconds are required. Treatment continues until no further secretions are expulsed and secretion related oxyhemoglobin desaturations are reversed. Use can be required as frequently as every 30 minutes around the clock during chest infections.

The use of mechanical insufflation-exsufflation (MI-E) via the upper airway can be effective for children as young as 11 months of age. Patients this young can become accustomed to MI-E and permit its effective use by not crying or closing the glottis. Between 2.5 and 5 years of age, most children become able to cooperate and cough on queue. Exsufflation-timed abdominal thrusts are also used for infants.

Whether via the upper airway or via indwelling airway tubes, routine airway suctioning misses the left main stem bronchus about 90% of the time.(17) MAC provides the same exsufflation flows in both left and right airways without the discomfort or airway trauma of tracheal suctioning. Patients prefer MAC to suctioning for comfort and effectiveness, and they find it less tiring. (18) Deep suctioning, whether via airway tube or via the upper airway, can be discontinued for most patients.

VC, pulmonary flow rates, and SpO_2 when abnormal improve immediately with clearing of airway secretions and mucus by MI-E.(19). An increase in VC of 15% to 42% was noted immediately following treatment in 67 patients with "obstructive dyspnea," and a 55% increase in VC was noted following MI-E in patients with neuromuscular

conditions (20). We have observed 15% to 400% (200 to 800 mL) improvements in VC and normalization of SpO_2 as MI-E eliminates airway mucus for ventilator-assisted NMD patients with chest infections. (21)

Of the three muscle groups required for effective coughing, MI-E can only take the place of the inspiratory and expiratory muscles. Thus, it cannot be used to avert tracheotomy very long if bulbar-innervated muscle function is inadequate to prevent airway collapse and continuous saliva aspiration as often becomes the case in advanced bulbar ALS. On the other hand, patients with completely intact bulbar muscle function, such as most ventilator users with traumatic tetraplegia, can usually air stack to volumes of 3 L or more, and, unless very scoliotic or obese, a properly delivered abdominal thrust can result in assisted CPF of 6 to 9 L/s. These flows should be more than adequate to clear the airways and prevent pneumonia and ARF without need for MAC. Thus, the patients who benefit most from MAC have moderately impaired bulbar muscle function that limits assisted CPF to less than 300 L/m. This is typical of most non-ALS NMD patients, especially those with Duchenne muscular dystrophy (DMD) who benefit greatly from MAC. (16) Patients with respiratory muscle weakness complicated by scoliosis and inability to capture the asymmetric diaphragm by abdominal thrusting also greatly benefit from MI-E.

Glossopharyngeal Breathing

Both inspiratory and, indirectly, expiratory muscle function can be assisted by GPB (22). GPB can provide an individual with weak inspiratory muscles and no VC or breathing tolerance with normal alveolar ventilation when not using a ventilator or in the event of sudden ventilator failure day or night. (22, 23) The technique involves the use of

the glottis to add to an inspiratory effort by pistoning (gulping) boluses of air into the lungs. The glottis closes with each "gulp." One breath usually consists of 6 to 9 gulps of 40 to 200 mL each. 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, (24) and numerous videos are available (25), the best of which was produced in 1999. (26)

Although severe oropharyngeal muscle weakness can limit the usefulness of GPB, we have managed 13 DMD ventilator users who had no breathing tolerance other than by GPB (27). Approximately 60% of ventilator users with no autonomous ability to breathe and good bulbar muscle function can use GPB and discontinue ventilator use for minutes to up to all day. (22, 28) GPB is rarely useful in the presence of an indwelling tracheostomy tube. The safety and versatility afforded by GPB are additional reasons to eliminate tracheostomy in favor of noninvasive aids.

Because of their generally intact bulbar musculature, high level spinal cord injury (SCI) patients are ideal candidates to master GPB for ventilator-free breathing and be decanulated to NIV. In some centers, these patients are decannulated to free them from the fear of ventilator failure or accidental ventilator disconnection.(22, 28)

Oximetry Monitoring and Feedback Protocol

For a hypercapnic patient with desaturation due to chronic alveolar hypoventilation or the patient being weaned from tracheostomy ventilation, introduction to and use of mouthpiece or nasal NIV is facilitated by oximetry feedback. A SpO_2 alarm set at 94% signals the patient to normal SpO_2 by taking deeper breaths and to maintain SpO_2 over 94% all day. (16) When no longer possible to achieve this by unassisted

breathing, it is done by mouth piece or nasal NIV. With time, the patient requires increasing periods of NIV to maintain normal SpO_2. In this manner, central ventilatory drive can be reset.

Continuous SpO_2 feedback is especially important during respiratory tract infections. The cough of infants and small children who can never sit is inadequate to prevent chest cold–triggered pneumonia and ARF. The patients use MAC for any dip in SpO_2 below 95%. When using NIV continuously, such dips are usually due to bronchial mucous plugging, and if not quickly cleared, atelectasis and pneumonia can quickly result. Thus, patients are instructed to use NIV and MAC to maintain normal SpO_2 to avert pneumonia, ARF, and hospitalization. For adults with infrequent chest colds, rapid access to MAC may be all that is necessary.

Invasive Ventilatory Support

The use of noninvasive aids can be contraindicated by the presence of: depressed cognitive function, orthopedic conditions interfering with noninvasive interface use, pulmonary disease necessitating high fiO_2, or uncontrolled seizures or substance abuse. (29) Also, the presence of a nasogastric tube can hamper the fitting of a nasal interface and the use of mouthpiece or nasal NIV by interfering with both soft palate closure of the pharynx and seal at the nose. Although tracheostomy ventilation can extend survival for NMD patients (30), morbidity and mortality outcomes are not as favorable as by noninvasive approaches. (31, 32) Tracheotomy is indicated for severe bulbar ALS patients (1), rarely if ever for DMD and SMA patients except for the occasional SMA type 1 patient. (33) Patients with DMD, even those who are continuously ventilator dependent on noninvasive NIV, can avoid hospitalizations and pulmonary morbidity and

mortality for decades and tracheotomy indefinitely when properly managed by using respiratory muscle aids. (16)

Long-Term Outcomes

SMA type 1 – We reported 17 SMA-1 patients with ventilation via tracheostomy tubes, mean age 78.2 (range 65–179) months, 25 of 27 lost all autonomous breathing ability immediately upon tracheotomy. None of the 21 who had not developed the ability to verbalize before undergoing tracheotomy did so after tracheotomy. On the other hand, 72 SMA-1 patients using NIV are alive at mean age 86.1 (range 13–196) months; 13 died at 52.3 (range 13–111) months. Sixty seven of the 75 could communicate verbally. Fifteen SMA-1 patients are now over age 10 and 6 over age 15 without tracheostomy tubes and despite requiring continuous NIV in most cases. (33) Others have also reported continuous NIV dependence for patients with SMA type 1. (34)

DMD - 101 of our nocturnal-only NIV users eventually became continuously NIV dependent for 7.4±6.1 years to 30.1±6.1 years of age with 56 patients still alive. Twenty-six of the 101 became continuously dependent without requiring hospitalization. Eight continuous tracheostomy ventilation users were decanulated to noninvasive NIV. Thirty-one consecutive "unweanable" intubated patients were extubated to NIV/MAC. Seven of our DMD patients have lived to over age 40 including four who have required NIV continuously for 28, 19, 21, and 24 years to ages 41, 44, 48, and 47. Others have also reported prolongation of life for DMD by continuous NIV. (35)

ALS – Of 176 of our ALS patients using nocturnal NIV, 109 or 42% of all went on to require continuous NIV for about 10 months before their SpO_2 baseline decreased below 95% because of saliva aspiration due to bulbar-innervated muscle impairment.

At the 69th Congress of the Mexican Society of Respirology and Thoracic Surgeons, 20 centers from 14 countries presented data on over 1500 SMA1, DMD, and ALS patients who required continuous ventilatory support without tracheostomy tubes. Four of the centers routinely extubated unweanable DMD patients so that none of their over 250 continuously ventilator dependent or any other patients has undergone tracheotomy.

For Extubation of Unweanable Patients

NMD-specific extubation criteria and a new extubation protocol were developed. Once meeting the criteria, oro or nasogastric tube was removed to facilitate post-extubation nasal NIV. The patient was then extubated directly to NIV on assist/control 800 to 1500 ml, rate 10-14/min in ambient air. The NIV was provided via a combination of nasal, oro-nasal, and mouth piece interfaces. Assisted CPF, CPF obtained by abdominal thrust following air stacking, were measured within 3 hours as the patient received full volume-cycled NIV support. Patients kept 15 mm angled mouth pieces accessible and weaned themselves, when possible, by taking fewer and fewer intermittent positive pressure ventilations as tolerated. Diurnal nasal NIV was used for those who could not secure the mouth piece. They used nasal or oronasal interfaces for night time ventilation. For episodes of SpO_2<95%: ventilator positive inspiratory pressure (PIP), interface or tubing air leakage, CO_2 retention, ventilator settings, and MAC were considered. Patients were then taught air stacking and manually assisted coughing. Then assisted CPF were measured.

The therapists, nurses, and in particular, the family and personal care attendants provided MAC via oro-nasal interfaces up to every 30 min until SpO_2 no longer dipped below 95% and the patients felt clear of secretions. In 7 cases, post-extubation oral intake was considered unsafe so open modified Stamm gastrostomies were performed under local anesthesia using NIV without complication.

Data were reported on 157 consecutive "unweanable" patients, 25 with SMA, 20 with DMD, 16 with ALS, 51 with other NMDs, 17 with spinal cord injury, and 11 with polio. Eighty-three who refused tracheostomies were transferred from other hospitals. They could not pass spontaneous breathing trials before or after extubation. Once SpO_2 was maintained ≥95% in ambient air they were extubated to continuous NIV and aggressive MAC. Extubation success was defined as not requiring re-intubation during the hospitalization. Before hospitalization 96 (61%) patients had no experience with NIV, 41 (26%) used it part-time, and 20 (13%) were continuously NIV dependent. First attempt protocol extubation success rate was 95% (149 patients). All 98 extubation attempts on patients with assisted CPF ≥ 160 L/m were successful. Six of 8 patients who initially failed extubation succeeded on subsequent attempts, so only two bular ALS paitents with no measurable assisted CPF underwent tracheotomy. (36)

For Decanulation of Unweanable Patients

In 1996 we reported the decanulation of 50 unweanable patients with neuromuscular weakness. (3) Earlier, in 1990 and 1991 we and others reported the routine decanulation of high level traumatic spinal cord injured patients to NIV.(23,28) The principles of decanulating unweanable are essentially the same as those for extubation. Any ventilator dependent patient whose bulbar-innervated musculature is

adequate such that saliva aspiration does not cause a continuous decrease in baseline SpO2 is a candidate for decanulation to NIV. Patients with tracheostomy tubes who had no VFBA with VCs of 250 ml or greater invariably developed VFBA following decanulation. Most weaned to nocturnal-only NIV within 3 weeks of decanulation. Tube removal also facilitated speech and swallowing. All decanulated patients preferred NIV to tracheostomy ventilation for convenience, speech, swallowing, cosmesis, comfort, safety, and preferred it overall. (37)