Physical medicine respiratory muscle aids to avert respiratory complications of pediatric chest wall and vertebral deformity and muscle dysfunction

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The purpose of this article was to describe the use of noninvasive inspiratory and expiratory muscle aids to prevent chest wall deformities including pectus excavatum, to prevent respiratory complications of vertebral surgery, to prevent acute and long-term ventilatory insufficiency and failure in children with paralytic disorders who develop these deformities, 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 20 centers in 14 countries reported over 1 500 spinal muscular atrophy type 1 (SMA1), Duchenne muscular dystrophy (DMD), and amyotrophic lateral sclerosis (ALS) patients who survived using continuous ventilatory support without tracheostomy tubes. Four of the centers routinely extubated unweanable DMD patients so that none of their over 250 such patients has undergone tracheotomy.

KEY WORDS: Rehabilitation centers - Vertebral artery - Respiration, artificial - Assisted coughing - Child.

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Clinical population

Chest wall and vertebral deformities and ventilatory failure can present at birth. Floppy, hypotonic newborns for whom neuromuscular disorders are identified almost invariably develop progressive vertebral deformities and ventilatory impairment later in childhood (Table I). Unless treated by noninvasive inspiratory muscle aids, paradoxical breathing in infants results in *pectus excavatum* and undergrowth of the lungs and chest walls. Older children develop vertebral deformities from paralytic and/or hypertonic conditions (Table II). The latter are most often associated with static encephalopathies, dystonias, progressive central nervous system, genetic, and generalized diseases like familial dysautonomia.

It has been reported that for every 10° of thoracic scoliosis there is about a 4% decrease in vital capacity (VC). However, in neuromuscular scoliosis, VC is usually much lower than what could be caused by scoliosis. Thus, correction of scoliosis usually does not result in any increase, but most often a decrease in VC because spinal fixation also fixes the ribs and prevents chest expansion. Any children with severe bulbar-innervated muscle dysfunction also develop respiratory complications due to airway protection impairment.

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TABLE I.—*Neuromuscular diseases of floppy infants in order of incidence.*

Spinal muscular atrophy
Congenital myopathies
Congenital muscular dystrophies
Congenital myotonic dystrophy
Congenital myasthenic syndrome
Acid maltase deficiency (Pompe's disease)
Debranching enzyme deficiency (type 3 glycogen storage disease)
Branching enzyme deficiency (type 4 glycogen storage disease)
Carnitine deficiency
Mitochondrial myopathies
Congenital peripheral neuropathies

Respiratory muscle aids

It is important to review the respiratory muscle groups and the methods for assisting/supporting them. There are three respiratory muscle groups: the inspiratory muscles, expiratory (predominantly abdominal and upper chest wall) muscles for coughing, and the bulbar-innervated muscles. While the inspiratory and expiratory muscles can be completely supported such that even patients with 0 mL of VC have used NIV for over 50 years without resort to tracheostomy, there are no effective noninvasive measures to assist bulbar-innervated muscle function. Thus, the only indication for tracheotomy in an unweanable patient is the aspiration of saliva to the degree that the oxyhemoglobin saturation decreases and remains below 95%.[1] This does not usually occur until after a patient entirely loses the ability to speak and swallow food. Such patients develop essentially irreversible upper airway obstruction and require tracheostomy tubes to protect the airway.

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 (noinvasive intermittent positive pressure ventilation or NIV) assists the inspiratory muscles. Continuous positive airway pressure (CPAP) does not assist ventilation and is not useful for patients with weak inspiratory muscles.

TABLE II.—*Neuromusculoskeletal and central nervous system conditions which warrant physical medicine respiratory interventions.*

Skeletal including arthrogryposis, congenital vertebral deformities, congenital pectus, excavatum, Marfan's syndrome
Muscular dystrophies
 — Dystrophinopathies
   Duchenne and Becker muscular dystrophies
 — Non-dystrophin related muscular dystrophies limb girdle, Emery Dreifuss, facioscapulohumeral, congenital, childhood autosomal recessive, oculopharyngeal, and myotonic dystrophy
Non dystrophic myopathies
 — Congenital and metabolic myopathies including mucopolysaccharidoses, mucolipidoses, homocystinuria, carnitine deficiency, myotubular myopathy mitochondrial myopathies, inflammatory myopathies such as polymyositis
Diseases of the myoneural junction such as myasthenia gravis, congenital myasthenic syndromes, mixed connective tissue disease
Myopathies of systemic disease such as carcinomatous myopathy,
Cachexia/anorexia nervosa, medication associated
Neurological Disorders
 — Spinal muscular atrophies
 — Motor neuron diseases
 — Poliomyelitis
 — Neuropathies such as polyneuropathy, Guillain Barré Syndrome
 — High level spinal cord injury and other myelopathies
 — Hereditary sensory motor neuropathies including familial hypertrophic interstitial
 — Myelopathies of anatomic, rheumatoid, infectious, spondylitic, vascular, traumatic, neoplastic, or idiopathic etiology
 — Tetraplegia associated with pancuronium bromide, botulism
 — Central nervous disorders including static encephalopathies, Arnold Chiari malformation, Syringomyelia, Myelomeningocele, Encephalitis, Familial dysautonomia, Down's syndrome, multiple sclerosis, Parkinson's disease, Friedreich's ataxia
 — Phrenic neuropathies associated with cardiac hypothermia, surgical or other trauma, radiation, phrenic electrostimulation, familial, paraneoplastic or infectious etiology, and with lupus erythematosus

Historical perspective

Physiatrists developed all aspects of noninvasive positive pressure respiratory muscles aids. Sanatoria were built from 1885 in the United States for tuberculosis patients until the discovery of streptomycin in 1943 when they either closed, became hospitals, or were turned into the first rehabilitation centers for poliomyelitis survivors (PPS).[2] Many such PPS had been "living" in iron lungs in local hospitals since contracting polio. By 1949 there were over 400 re-

spiratory PPS in custodial care in over 100 U. S. hospitals.[2] Since hospital charges were high, patients were moved to 16 regional former sanatoria in the United States beginning in 1948.[2] They eventually managed 15 to 160 patients in each.[2] Pulmonologists and respiratory therapists did not exist at the time. The patients' and their families were taught by nurses and physical therapists how to use respiratory physical medicine aids to ventilate lungs and eliminate airway secretions and became the key participants in providing these aids. The 16 body ventilator units developed in the United States included one in Goldwater Memorial Hospital in Manhattan and one at Rancho Los Amigos National Rehabilitation Center in Downey, California.[2, 3] There, at Rancho in 1948, Dr. Clarence Wilding Dail at Rancho observed and reported glossopharyngeal breathing (GPB) by 15 PPS.[4] Dail reported that otherwise iron lung dependent patients could leave the iron lung for hours and maintain their breathing by GPB.[4] He and his colleague Dr. John E. Affeldt began instructing patients in GPB and developed a video tape and manual on it in 1954.[5]

At a poliomyelitis conference in 1953 Dr. John E. Affeldt, Rancho Medical Director from 1957 to 1964, wrote: "There is just one point I would like to mention, which has just come along which, I think, makes it (NIV) even more feasible. It is so simple that why it wasn't thought of long ago I don't know. Actually, some of our physical therapists, in struggling with the patients, noticed that they could simply take the positive pressure attachment, apply a small plastic mouthpiece [...], and allow that to hang in the patient's mouth […] if the patient is on the rocking bed and has a zero vital capacity, if you want to stop the bed for any reason […] you can simply attach this, hang it by the patient, he grips it by his lips, and thus it allows for the excess to blow off which he does not want. It works very well. We even had one patient who has no breathing ability who has fallen asleep and been adequately ventilated by this procedure, so that it appears to work very well, and I think does away with a lot of complications of difficulty of using (tracheostomy) positive pressure. You just hang it by the patients and they grip it with their lips, when they want it, and when they don't want it, they let go of it. It is just too simple".[6] Thus, Affeldt gave the first description of mouth piece NIV.

Dail and Affeldt were pioneers in the field of physical medicine and rehabilitation (PM&R). Dail was one of the first 91 physicians certified by the American Board of PM&R in 1947 and served as President of the American Academy of Physical Medicine and Rehabilitation 1959-60. Affeldt was President of the American Congress of Rehabilitation 1976-7. By 1952 continuously ventilator dependent PPS were being discharged home using chest shells, iron lungs, and intermittent positive pressure ventilators (IAPVs or "pneumobelts") including 81 of the at least 500 continuously body ventilator dependent patients in the US from 1952-1955.[2]

It was physiatrist Mark L. Splaingard *et al.*, however, who used body ventilators for those with non-polio neuromuscular disorders, describing 15 unweanable body ventilator users among the 30 with muscular dystrophy and spinal muscular atrophy going home between 1962 and 1981.[7] By 1956, 92% of continuously ventilator dependent individuals were being discharged home.[8] All that was available for mouth piece NIV, however, was using the exhaust of vacuum cleaners with water traps to limit pressures to about 40 cm H_2O "for periodic (lung) overdistension" and glossopharyngeal breathing.[8, 9] Once "portable" positive pressure ventilators became available in 1956, mouthpiece NIV became routine for daytime ventilatory support both at Goldwater Memorial Hospital and at Rancho.[8, 10] Its use was only first reported in a peer-reviewed medical journal by physiatrists in 1973.[11] In this paper, physiatrists Aberion, Alba, Lee, and Solomon reported its use along with that of iron lungs, rocking bed, chest shells, and the IAPV for DMD patients.[11]

Dr. Augusta Alba, board certified in physical medicine and rehabilitation (PM&R) in 1959 became director of the Goldwater Memorial Hospital ventilator unit in 1956.[10] She encouraged patients to use mouthpiece NIV for daytime support.[10] She soon discovered that mouthpiece NIV users would nap without the mouthpieces falling out of their mouths.[10] This was remarkable because many of these patients had no autonomous breathing ability; their ventilators did not have alarms; there was only the tongue to keep the mouthpiece in the mouth during sleep; and they had little or no limb function to put the mouthpieces back if they should fall out.[10] In 1964 she permitted patients to use mouthpiece NIV overnight rather than return to body ventilators.[10] The Bennett lipseal in 1968 kept the mouthpiece securely in place during sleep allowing Dr. Alba to convert most of her sleep body ventilator

users to mouthpiece/lipseal NIV. Bach, PM&R board certified in 1986, and Alba published 4 000 patient-years of ventilatory support by mouthpiece NIV for a combination of sleep and daytime NIV for 257 ventilator users in 1993.[12]

From 1968 until nasal NIV was described in 1987,[13-15] mouthpiece/Lipseal NIV was the only method of NIV.[10] In the meantime in 1979 physiatrists Michael Alexander and Ernest Johnson *et al.* published the first paper on long-term NIV support for 10 DMD patients and first recognized that tracheostomy could be avoided for them indefinitely.[16] They also reported that ventilator dependent DMD patients with tracheostomies could be decanulated and rely on around-the-clock body ventilator and mouth piece NIV use along with manually assisted coughing, that is, deep lung insufflation followed by abdominal thrust.[16] Decanulation of other ventilator dependent patients including high level tetraplegics was not to be repeated until reported by Bach *et al.* in 1990.[17, 18]

Despite ventilatory support by body ventilator use for decades, with the advent of the oximeter in 1979, Bach and Penek reported that negative pressure body ventilator use during sleep resulted in severe obstructive sleep apneas.[19] They switched 40 body ventilator users to NIV, thereby correcting the sleep breathing disorder, normalizing oxyhemoglobin saturation, and alleviating symptoms of CO_2 retention, as well as hypertension for five of the patients.[19] Although due to airway collapse during sleep, body ventilator use is less effective than NIV, the IAPV, a body ventilator used for daytime support, was still useful. In 1991 physiatrists Bach and Alba reported 940 patient-years of IAPV use by 52 continuously ventilator dependent patients with neuromuscular ventilatory failure.[20]

The next NIV milestone was the invention of nasal NIV. Bach was employed by French physiatrist Yves Rideau of the PM&R department of a Poitiers, France medical school to institute NIV from 1981 to 1983. Upon arrival in June of 1981, Rideau proposed to use NIV *via* nasal access rather than mouthpiece for daytime NIV and to use it prophylactically during sleep for young, asymptomatic DMD patients in a futile attempt to decrease the rate of loss of VC in this condition. Bach inserted Foley catheters into his nostrils, inflated the cuffs, and attached himself to a ventilator in late 1981. Seeing that air entered the lungs alright, it

was used by DMD patients for deep daytime insufflations as well as during sleep and was first published in the Ph.D. thesis of physiatrist Anne Delaubier in 1984.[21] Nasal NIV was first used for continuous ventilatory support as an alternative to airway intubation by a multiple sclerosis patient with 100 ml of VC in 1984 and published by physiatrists Bach, Alba, Mosher, and Delaubier in 1987.[13] Bach then switched Lipseal NIV users with no measurable VC to using nasal NIV during sleep and realized that nasal NIV could provide full ventilatory support during sleep as well. Nasal "CPAP" masks became available in 1984 and were used for NIV delivery,[22-24] and Bach *et al.* also described the customization of nasal interfaces in 1989.[25] These methods were introduced into Japan in 1991 by physiatrist Dr. Susume Ohtake.[26]

It is now increasingly recognized that NIV can be used indefinitely by even patients with no measurable VC. Children with spinal muscular atrophy (SMA) type 1, continuously NIV supported for 15 years or more, are now approaching 18 years of age.[27] Fifteen hundred continuous NIV users with DMD, SMA, or amyotrophic lateral sclerosis from 20 centers in 14 countries were reported at the 69th Congress of the Mexican Society of Respirologists and Thoracic Surgeons on April 6-9, 2010 by a group that included 9 physiatrists. Over 200 continuously ventilator dependent NIV users, intubated for episodes of acute respiratory failure, were extubated back to continuous NIV rather than undergo tracheostomy. In 2010 Bach reported the systematic extubation of 157 "unweanable" patients to NIV rather than resort to tracheostomy, thereby permitting them to return home rather than to a lifetime of institutionalization for most cases.[28]

The most important complimentary interventions that permit the long-term use of NIV include air stacking which was first used by Dr. Alba and ultimately reported by physiatrists Kang and Bach,[29-31] manually assisted coughing first reported by physiatrist Arthur A. Siebens, PM&R board certified in 1984,[32] extubation and decanulation of unweanable patients reported by Bach *et al.*,[33, 34] and mechanically assisted coughing (MAC).[35-37] The latter deserves special note because of its importance in allowing NIV users to survive intercurrent respiratory tract infections without developing respiratory failure and then undergoing tracheotomy.
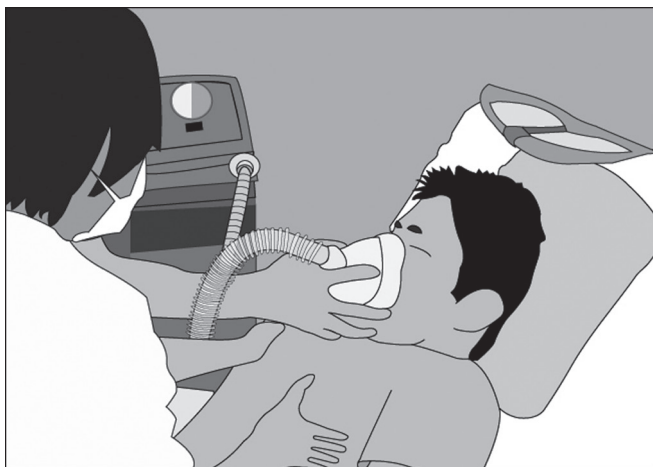
Figure 1.—Mechanically assisted coughing: mechanical Insufflation-Exsufflation with chest compression at time of expiration.

In May of 1953 Dr. John Affeldt noted that he had had his patients try many methods of mechanically assisted coughing including the suction of vacuum cleaners, manual chest and abdominal compression, and manipulating iron lung pressure delivery against a closed glottis to increase cough flows.[35, 36] Jack Emerson noted that it makes more sense to provide positive than negative pressure to the airways than to use iron lung "cough chambers".[35, 36] The Cof-lator™ came onto the market in 1954 but the age of the tracheostomy had arrived no one apparently ever used the Cof-flator™ *via* the tube to discover how effective and comfortable it can be so it went off the market.[35, 36] PPS and other NIV users who had vintage devices, however, would not give them up because they knew that not having one during a respiratory tract infection meant certain respiratory failure and intubation.[35, 36] In the late 1980s several patients wrote open letters to newspapers asking that the device be put back onto the market.[37, 38] Dr. Bach solicited every ventilator manufacturer to redevelop the device. He obtained a Food And Drug Administration Orphan Products Development Grant in 1990 to re-develop and study a cough assisting device and published a paper on such a device's pressure-flow characteristics in 1993.[37, 38] Only Jack H. Emerson, the manufacturer of iron lungs since 1931, re-manufacturered it and brought it back onto the market in February 1993 as the In-exsufflator™ and later, the CoughAssist™ (Figure 1).

Thus, instead of letting patients develop certain respiratory failure, physiatrists developed virtually every aspect of supporting patients noninvasively, including NIV, MAC, and all complementary interventions to prevent respiratory failure, hospitalization, institutionalization, and resort to invasive airway tubes, and to extubate and decanulated patients with the tubes to restore quality of life. It is unlikely that "sleep physicians" or other physicians who, to this day, limit themselves to using CPAP or bi-level positive airway pressure (bilevel-PAP) for patients with neuromuscular weakness would have ever conceived of ventilatory support by NIV nor would it now be used anywhere if not for physiatrists.[39]

Respiratory pathophysiology

Respiratory impairment results from either primarily lung/airways diseases, in which case pulmonary function testing can be indicated and supplemental oxygen and bronchodilators beneficial, or it results from complications of respiratory muscle impairment. The former is characterized by hypoxia in the presence of eucapnia or hypocapnia until an exacerbation causes acute respiratory failure (ARF). The latter is characterized by hypercapnia and hypoxia caused by hypoventilation or respiratory insufficiency/failure due to an ineffective cough. The former is respiratory insufficiency/failure whereas the latter may be ventilatory insufficiency/failure. Unfortunately, physicians rarely distinguish between the two, referring to both, as well as evaluating and treating both, as respiratory insufficiency/failure. This results in needless morbidity and mortality, not to mention cost and decreased quality of life. Symptomatic hypercapnic patients benefit from NIV for at least part of the day and, more often overnight. With progressive inspiratory muscle weakness, ventilator-free breathing ability is eventually lost. Airway mucous plugging due to an ineffective cough is reversible by using expiratory (cough) aids.

Ventilatory insufficiency/failure can be nocturnal only and result from diaphragm dysfunction with the patient unable to breathe when supine; it can be from total inspiratory muscle failure; it can result from inadequate central ventilatory drive, or it can result from severe obesity or chest wall restriction.

TABLE III.—*Patient evaluation.*

Family history of pulmonary diseases

Symptom progression and impact on function

Exacerbation and hospitalization history

Appetite, nutritional status and weight changes

Medications, substance abuse

Physical examination for hyperresonant chest, poor breath and Cardiac sounds

Hemoglobin/hematocrit, sedimentation rate, C-reactive protein, white cell count

Radiographic assessment of (low, flattened) diaphragm, (long, narrow) heart shadow, retrosternal translucency, narrowing of peripheral pulmonary vessels

$PaCO_2$ and PaO_2 and diffusion capacities (decreased in emphysema, normal in bronchitis)

Pulmonary function tests for air and mucus trapping

Low maximum mid-expiratory flow rates and increased mid-expiratory times, normal or increased lung compliance and increased flow work, increased residual volume and total lung capacity

Clinical exercise testing

A 3-, 6-, or 12-minute walk

Assessment of the ventilatory anaerobic threshold and maximum exercise tolerance for a precise exercise regimen

Many patients with ventilatory insufficiency survive for years without ventilator use at the cost of orthopnea and increasing hypercapnia with its associated symptoms and dangers and a compensatory metabolic alkalosis that depresses central ventilatory drive. The alkalosis allows the brain to accommodate to hypercapnia without overt symptoms of acute ventilatory failure. Hypercapnic patients not using NIV, and especially those receiving supplemental oxygen, develop increasingly severe hypercapnia that eventually results in coma from carbon dioxide narcosis and ventilatory arrest. When symptomatic, hypercapnic patients are properly treated with NIV, blood gasses normalize, and symptoms and alkalosis resolve as the kidneys excrete excess bicarbonate ions. Because of the need to take bigger breaths to maintain normal $PaCO_2$ and blood pH when discontinuing NIV in the morning, dyspnea can necessitate increasing periods of daytime NIV until patients require NIV continuously. Some patients with ventilatory muscle failure and no measurable VC with their respiratory muscles use only nocturnal aid and rely on glossopharyngeal breathing (GPB) to ventilate their lungs during daytime hours.

Patient evaluation

Patients with diminished ventilatory reserve who are able to walk commonly complain of exertional dyspnea (Table III). Eventually, morning headaches, fatigue, sleep disturbances, and hypersomnolence develop.[40] 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 requires four items: a spirometer, peak flow meter, non-invasive CO_2 monitor (transcutaneous or 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 the difference is greater than 20%, orthopnea is common and 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 maximum lung volumes 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.[30] A nasal interface, lipseal, or oro-nasal interface 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, USA) (Figure 2). CPF of 160 L/m are the minimum needed to cough effectively,[34] and this is the best indicator for tracheostomy tube removal irrespective of remaining pulmonary function for older children and adults. Patients with VCs less than 1 500 mL have assisted CPF measured from a maximally stacked volume of air and with an abdominal thrust and or thoracic compression delivered simultaneously with glot-

Figure 2.—Measuring cough peak flow with peak flow meter. Cough peak flows increase from ineffective levels to effective levels by manually assisted coughing with maximally insufflating the patient and then providing a chest compression.

tic opening (Figure 2).[30] Coughing from a deep air stacked volume with a concomitantly applied abdominal thrust and or thoracic compression 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.[41] Non-invasive continuous blood gas monitoring, including transcutaneous CO_2 monitoring or 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 transcutaneous pCO_2 monitor or capnograph, which measures end-tidal pCO_2, must be capable of summarizing the data.[26, 42] 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 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 coughing and inadequate airway secretion management, any patient finding that NIV use is more burdensome than

symptoms of ventilatory insufficiency is told to discontinue NIV and return for a re-evaluation in three to six 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 prescribed.[43] Obesity-hypoventilation patients are treated with nocturnal ventilatory support, as are neuromuscular disease (NMD) patients, and not with CPAP. Polysomnography is not indicated for patients with decreased VC (especially with 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 children with vertebral deformity and NMD 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 and to avoid hospitalizations, and prolong survival without resorting to tracheotomy. Unweanable intubated and canuled patients can be extubated and decanulated to NIV and 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 cannot 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.[31]

Figure 3.—Air stacking using a manual resuscitator via a mouthpiece.

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 undergo tracheotomy.[29] Patients who cannot close the glottis and, therefore, cannot air stack, must be passively insufflated using a CoughAssist™ (Philips Respironics, Amsterdam, the Netherlands), a pressure-cycling ventilator at pressures of 40 to 70 cm H_2O, or a manual resuscitator with the exhalation valve blocked. The maximum passive insufflation volume can be termed the "Lung Insufflation Capacity" or LIC.[44]

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 1 131±744 mL, 1712±926 mL, and 2069±867 mL, respectively.[44] 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 predict-ed 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 and small children with SMA and others with infantile NMDs who have paradoxical chest-wall movement require nocturnal NIV to prevent pectus excavatum and promote lung growth as well as for ventilatory muscle rest.[45] 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 small child's breathing. Children can become cooperative with deep insufflation therapy by 14 to 30 months of age (Figure 3).

Goal two: maintain normal alveolar ventilation by inspiratory muscle assistance

Negative pressure body ventilators cause obstructive apneas and become less effective with age and decreasing pulmonary compliance.[23] The intermittent abdominal pressure ventilator (IAPV) or "Exsufflation Belt™" (Philips Respironics. Amsterdam, the Netherlands), however, 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° 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.[20]

Non-invasive intermittent positive pressure ventilation

IPPV can be non-invasively 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 insuffla-

tion leakage during sleep,[24, 46] thus, supplemental oxygen and sedatives can render NIV ineffective. NIV can be introduced in the clinic or home setting.

There are numerous commercially available nasal interfaces (CPAP masks). When using "CPAP masks" on a ventilator circuit with an exhalation valve, the exhalation holes of the interface need to be plugged or covered and mask leak eliminated. Insufflation leakage can also be decreased 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. 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 used for daytime ventilatory support. Some patients keep the 15 mm angled mouthpiece between their teeth all day.[47] 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 300 to 1 500 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 posteriocaudally 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.[48]

Nasal NIV is also indicated for infants and for those who cannot grab or retain a mouth piece because of oral muscle weakness, inadequate jaw opening, or insufficient neck movement. Continuous mouth piece/nasal NIV is a desirable alternative to tracheostomy.[24] Nasal IPPV 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 IPPV.[49] This can be reduced by warming the inspired air to body temperature and humidifying it using a hot-water bath humidifier.[49] Decongestants can also relieve sinus irritation and nasal congestion. Other than perhaps for an uncontrollable seizure disorder, there are no absolute 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 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 1 000 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

With the higher lung volumes by air stacking, assisted CPF of 4.3±1.7 L/s were obtained by comparison with 2.5±2 L/s unassisted in patients over 12 years of age.[31] 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 1 647.6 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.[50] In older children, 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.

The availability of reference levels for CPF in the pediatric population (healthy children ages 4-18 yrs)

may be helpful for establishing the risk of acute respiratory complications for young patients with weak coughs, particularly those with vertebral deformity.[51]

MAC is the combination of the use of mechanical insufflation-exsufflation (MI-E, using the 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. MAC can be used for infants and is often felt to be effective by parents and therapists. However, its use must be timed to the child's breathing cycle. Its use *via* the upper airway can certainly be effective for children as young as 11 months of age when they become accustomed to it 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.[52] 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.[53] 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 se-

cretions and mucus by MI-E.[54] 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 NMD.[55] 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.[36]

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 occasionally becomes the case in SMA type 1. On the other hand, patients with completely intact bulbar muscle function, such as most ventilator users with traumatic tetraplegia over age 12, 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 children with SMA, DMD, and other NMDs.[50] Patients with respiratory muscle weakness complicated by scoliosis and inability to capture the asymmetric diaphragm by abdominal thrusting or chest compression can also greatly benefit from MI-E.

Glossopharyngeal breathing

Both inspiratory and, indirectly, expiratory muscle function can be assisted by GPB.[56] 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.[18, 56] 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 train-

TABLE IV.—*Management of patients with spinal cord injury.*

Level*	VC ml	Neck function Bulbar muscles**	Daytime	Nocturnal
Above C1	0	Inadequate/inadequate	TIPPV	TIPPV
C2–C3	<200	Adequate/inadequate	EPR_	N/MIPPV
Below C2	>200	Adequate/adequate	MIPPV/IAPV	N/MIPPV

*Motor levels; **Adequate neck function involves sufficient oral and neck muscular control to rotate, flex, and extend the neck to grab and use a mouthpiece for IPPV; adequate bulbar function to prevent aspiration of saliva to the degree that the SpO_2 baseline decreases below 95%. EPR: electrophrenic pacing; IAPV: intermittent abdominal pressure ventilator; MIPPV: mouthpiece intermittent positive pressure ventilation; NIPPV: nasal intermittent positive pressure ventilation; TIPPV: tracheostomy intermittent positive pressure ventilation.

ing manual and numerous videos are available,[5] the best of which was produced in 1999.[57]

Although severe oropharyngeal muscle weakness can limit the usefulness of GPB, we have reported 13 DMD ventilator users who had no breathing tolerance other than by GPB.[58] 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.[17, 56] 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 decanuled to NIV. In some centers, these patients are decannulated to free them from the fear of ventilator failure or accidental ventilator disconnection (Table IV).[17, 56]

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.[50] 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 older children with infrequent chest colds, rapid access to MAC may be all that is necessary.

Preparation for surgery, and extubation despite failure to breathe spontaneously kyphoscoliosis and ventilatory dysfunction

As previously noted, idiopathic kyphoscoliosis reduces VC and respiratory reserve independently of any concomitant muscle weakness. For patients with scoliosis due to NMD, however, decrease in VC and respiratory reserve are due almost entirely to respiratory muscle weakness. This is clear because surgery to correct scoliosis generally decreases rather than increases VC and no improvement occurs in the subsequent rate of loss of VC.[42] Patients with kyphoscoliosis also develop ventilation-perfusion mismatching and have a higher incidence of obstructive sleep apneas when the skeletal deformities involve the upper cervical region and, therefore, the hypopharynx.[59, 60] Although not necessarily decreasing VC, scoliosis can exacerbate the loss of pulmonary compliance which, in turn, increases the work of breathing.

Optimization of preoperative nutritional status may involve the use of mouthpiece IPPV for deep breaths between swallows to permit sufficient time to swallow safely. This often results in improved nutritional intake and weight gain.[42]

Respiratory management for scoliosis surgery in DMD

INDICATION AND TIME OF SPINAL SURGERY

In recent years, early prophylactic spinal instrumentation has been recommended in Europe and in the United States as the treatment of choice for the spinal deformities and ensuing seating problems.[61]

Overall, sublaminar, hybrid and pedicle screw instrumentation system constructs appear to provide and maintain an optimal degree of correction long term but the advantages of lesser blood loss and surgical time without the need for pelvic fixation seem to favor the pedicle screw system.[62]

Preexisting restrictive lung disease and muscle weakness place a patient with SMA or DMD at risk for perioperative pulmonary complications that can result in the failure to pass spontaneous breathing trials (SBTs), conventionally needed for extubation without tracheotomy. Therefore, early spinal surgery had been recommended before further respiratory muscle weakness results in the VC decreasing below 40%.[63] We attempt to determine those patients in whom spinal deformity will rapidly progress and to whom surgical intervention should be applied as early as possible. In 1996 we suggested that when the VC plateau occurs before age 14 years of age or when the value of the VC plateau is less than 1 900 mL, surgical treatment for the spinal deformity should be considered earliest.[64] More recently, with preoperative training in NIV and MAC and extubation to full-setting NIV and MAC it is clear that scoliosis surgery can be done without respiratory complications even for patients with little or no VC.[65] Therefore, we now recommend surgery when the Cobb angle approaches 40°, or sometimes 30° if rapidly progressive, irrespective of pulmonary function.

ANESTHESIA AND SEDATION

The risks related to anesthesia and sedation in DMD patients include potentially fatal reactions to inhaled anesthetics and certain muscle relaxants, upper airway obstruction, hypoventilation, atelectasis, congestive heart failure, cardiac dysrhythmias, respiratory failure, and difficulty weaning from mechanical ventilation.[66] Therefore, the multidisciplinary attention of specialists in respiratory, cardiac, gastrointestinal, and anesthetic management is important before providing general anesthesia or procedural sedation.[66]

Accordingly, preoperative pulmonary assessment including VC and CPF should be performed.[66] If FVC is less than 30%, patients are trained in NIV.[66] If CPF are less than 270 L/min, they are also trained in assisted coughing and MAC.[66]

Postoperatively, patients are extubated to full setting NIV and the family present to use MAC up to every 30 minutes as needed to maintain the SpO_2

greater than or equal to 95%. SpO_2 should be monitored continuously until cardiopulmonary status remains stable and SpO_2 over 94%. Whenever possible, carbon dioxide levels are monitored by continuous or frequent intermittent transcutaneous or end-tidal pCO_2. If SpO_2 decreases below 95% carbon dioxide levels are assessed for hypoventilation etiology, and if atelectasis or airway secretions are the cause, they are treated by MAC. Supplemental oxygen therapy should be used with caution because it does not treat the underlying problem (*e.g.*, hypoventilation, atelectasis, or airway secretions) and oxygen therapy impairs central respiratory drive and can increase insufflation leakage out of the nose and mouth, especially during sleep.[10]

Invasive ventilatory support

The use of non-invasive aids can be ineffective or contraindicated by the presence of: depressed cognitive function, orthopedic conditions interfering with non-invasive interface use, pulmonary disease necessitating high FiO_2, or uncontrolled seizures or substance abuse.[67] 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,[36] morbidity and mortality outcomes are not as favorable as by non-invasive approaches and tracheostomy often necessitates lifetime institutionalization.[36, 68] Tracheotomy is rarely indicated for DMD and SMA patients except for the occasional SMA type 1 patient who aspirates too much to keep SpO_2 over 94%.[27] Patients with SMA and DMD, even those who are continuously ventilator dependent on NIV, can avoid hospitalizations and pulmonary morbidity and mortality for decades and tracheotomy indefinitely when properly managed by using respiratory muscle aids.[50, 69]

Brace and seating systems for neuromuscular scoliosis

It has been suggested that up to 37% of patients with DMD do not develop scoliosis by 16 years of age.[70] Considering the recently reported decreased

incidence of scoliosis in DMD patients treated with corticosteroids including deflazacort,[71] an approach to spinal surgery tailored only to those children needing it would be desirable.[72] The improved long-term prognosis of individuals with DMD who are now expected to survive into their 30s and beyond [47, 73] could further support the need for surgical stabilization of spine to maintain long-term seating comfort.

Since the kyphoscoliosis that develops in children with NMD is due to paraspinal muscle weakness and the inability of weak paraspinal muscles to hold up the spine, thoracolumbar bracing can not correct scoliosis or decrease the rate of scoliosis progression. In addition, resort to bracing often delays surgical correction to the point where pulmonary function decreases so much that many centers no longer recommend surgery. Although not affecting the scoliosis itself, bracing may help to maintain reversibility of the scoliotic curve for better surgical correction and it is indicated when the patient is too young for surgical correction. Some centers, however, continue to use bracing for DMD if the Cobb angle is greater than 20°. Bracing is certainly warranted for children with SMA below the age of 6 to allow a longer time for the spine to grow before resorting to scoliosis correction surgery.

Polypropylene moulded thoracolumbar spinal orthoses (TLSOs) can be offered.[72, 74] Patients are advised to wear their TLSOs whenever sitting.[72] We support the trunk while minimizing abdominal restriction that might impede breathing.[26] In Kinali's study, approximately a quarter of DMD patients (28/123, 23%) did not have any scoliosis during their first to second decades, or only no more than 30° scoliosis at age 17.[72] And 16 out of 123 (13%) patients who refused surgery or for whom surgery was thought to be contraindicated because of low VC or cardiac dysfunction had mild to moderate scoliosis (31-50°) that was managed conservatively by seating modifications and supports.[72] Nine patients developed a curvature of at least 50° but were treated conservatively as sitting posture was maintained in brace.[72] A specialized team including a physiatrist, physical therapist, and occupational therapist carefully monitored the patients and introduced an appropriate seating system on the patient's motorized wheelchair that minimized work of breathing while permitting comfortable breathing and seating comfort.[26]

Long-term outcomes

SMA type 1

Of 27 SMA1 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 were 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 SMA1 patients are now over age 10 and 6 over age 15 without tracheostomy tubes and despite requiring continuous NIV, in many cases for over 15 years.[27] Others have also reported continuous NIV dependence for patients with SMA type 1.[75]

DMD

One hundred ninety of our nocturnal-only NIV users eventually became continuously NIV dependent for 8±6 years to 30.4±6.2 years of age with 90 patients still alive. At least 26 became continuously NIV dependent without requiring hospitalization. Eight continuous tracheostomy IPPV users were decanulated to NIV. Forty-two 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.[76]

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 an extubation protocol were developed (Table V). Once meeting the criteria, any oro or nasogastric tube is removed to facilitate post-extubation nasal NIV. The

TABLE V.—*Extubation criteria for unweanable ventilator dependent patients.*

- Afebrile and normal white blood cell count
- No ventilator-free breathing tolerance with 7 cm pressure support in ambient air
- $PaCO_2$ 40 mm Hg or less at peak inspiratory pressures less than 35 cm H_2O on full ventilatory support as needed
- Oxyhemoglobin saturation (SpO_2) ≥ 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

patient is then extubated directly to NIV on assist/control 800 to 1 500 mL, rate 10-14/min in ambient air. The NIV is provided *via* a combination of nasal, oro-nasal, and mouth piece interfaces. Assisted CPF, CPF obtained by abdominal thrust and chest compression following air stacking, are measured as the patient receives full volume-cycled NIV support. Patients keep 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 is used for those who can not secure the mouth piece. Nasal or oronasal interfaces are used for ventilation during sleep. For episodes of $SpO_2<95\%$: ventilator positive inspiratory pressure (PIP), interface or tubing air leakage, CO_2 retention, ventilator settings, and MAC are considered. Patients are then taught air stacking and manually assisted coughing. The therapists, nurses, and in particular, the family and personal care attendants provide MAC *via* oro-nasal interfaces up to every 30 min until SpO_2 no longer dipping below 95% and the patients feel clear of secretions.

Data were reported on 157 consecutive "unweanable" patients, 25 with SMA, 20 with DMD, 78 with other NMDs, and 17 with spinal cord injury. Eighty-three who refused tracheostomies were transferred from other hospitals after failing extubation there. They could not pass SBTs 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. Only two bulbar ALS patients with no measurable assisted CPF underwent tracheotomy.[28]

For decanulation of unweanable patients

In 1996 we reported the decanulation of 50 unweanable patients with neuromuscular weakness.[33] Earlier, in 1990 and 1991 we and others reported the routine decanulation of high level traumatic spinal cord injured patients to NIV.[17, 18] 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 SpO_2 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 three 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.[77]

The "lifeline" for child's life using NIV

Families' lives change with the decision to place their child with neuromuscular disorders on mechanical ventilation.[78] Despite becoming expert caregivers, the parents experience a sense of loss and uncertainty. Those who perceive insufficient support feel the weight of responsibility as sole care providers for their child.[78] More support and advocacy by health care professionals and the community are needed [77, 78] to support the parents with their decision.

Although "multidisciplinary teams" are commonly called for to care for neuromuscular disease patients,[61, 72, 77] they are expensive, the care usually disjointed, and the centers calling for them rarely if ever have any single person with the expertise to

spare their patients from developing acute respiratory failure or undergoing tracheotomy. Such "teams" rarely, if ever, include anyone who understands what should be optimal care for all of the various disciplines. For example, without the referring physician understanding pathokinesiology and optimal lower limb orthopedic management, he/she will not find an orthopedic surgeon expert in preventing the lower limb contractures that can prolong brace-free ambulation or the spinal surgical team that can repair the scoliosis and extubate an unweanable patient without resort to tracheostomy. Thus, a patient is far better off with one physician with knowledge of the interventions and outcomes appropriate to optimal multidisciplinary care than seeing clinicians in all disciplines who have no special expertise in state-of-the-art neuromusculoskeletal management. Thus, one team leader needs to coordinate care and for that, he needs to have access to state-of-the-art literature on the management of these conditions. Ideally, this material should be available from a single source, that is, the only book ever published in English on the management of patients with neuromuscular disorders.[42]

Conclusions

Paralytic scoliotic syndromes are associated with hypoventilation and respiratory complications in the pediatric population. A simple evaluation to assess a child's respiratory muscle function rather than a full battery of pulmonary function tests designed for obstructive/intrinsic lung disease, and the application of pressures to the body and airways to support inspiratory and expiratory muscle function rather than supplemental oxygen therapy and bronchodilators can permit many progressively weakening children with vertebral deformity to avoid ARF. Those who do develop ARF, are intubated, and who can not pass SBTs can, nevertheless, be extubated to full-setting NIV and MAC and, thereby, avert tracheotomy provided that glottic function is sufficient to avoid secretion aspiration to the extent that baseline SpO_2 remains below 95%. Thus, an entirely different evaluation and treatment paradigm is required for the optimal and humane management of children with respiratory muscle weakness and vertebral deformity rather than lung disease.

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