

Abstract

Most patients with impairment of pulmonary function can be differentiated into those who have primarily oxygenation impairment with hypoxia due to predominantly intrinsic lung/airways disease and for whom hypercapnia is an end stage event, and those with lung ventilation impairment on the basis of respiratory muscle weakness for whom hypercapnia causes hypoxia. This distinction is important because, although many patients in the former category have been described to benefit from noninvasive ventilation in the acute care setting, long term use is more controversial. Patients with primarily ventilatory impairment, on the other hand, can benefit from the use of both inspiratory and expiratory muscle aids; and often avoid having any episodes of respiratory failure despite total respiratory muscle paralysis, do not require tracheostomy, and have excellent prognoses with long term home mechanical ventilation.

Ventilatory muscle failure is defined by the inability of the inspiratory and expiratory muscles to sustain one's respiration without resort to ventilator use. Patients with ventilatory muscle failure do not have unlimited breathing tolerance and require ventilatory support and other 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 most important inspiratory aid is to receive air under pressure when one inhales (intermittent positive pressure ventilation or IPPV). The most important expiratory aid is to have a negative pressure (vacuum) applied to the airway via the nose and mouth when one coughs along with a manual thrust to the belly to further increase cough flows. Illness and death in people with generalized weak-

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kness, such as patients with neuromuscular disease and high spinal cord injury, is almost always due to respiratory difficulty that occurs because of a weak cough. Breathing (inspiratory), expiratory, and throat (bulbar) muscles are needed for effective coughing. The latter are predominantly the abdominal muscles. Clearing airway secretions can

be a continual problem but it most often occurs during chest infections.

The following review will describe the most important aspects of pulmonary rehabilitation in patients with muscle weakness /paralysis and its main goals of respiratory muscle substitution to avoid ventilatory failure and promote quality of life in these patients.

Introduction

Ventilatory impairment is either due to central hypoventilation or respiratory muscle dysfunction in neuromuscular disease (NMD). Dysfunction results, most commonly, from weakness, contracture, or myotonia. As for the skeletal muscles, it is both the combination of weakness and contractures that result in dysfunction and this causes respiratory morbidity and mortality when untreated.

A typical example of ventilatory insufficiency is that of the hypercapnic patient with Duchenne muscular dystrophy (DMD) who has normal SpO₂ when awake and who is minimally symptomatic with little or no ventilator use. Patients symptomatic from ventilatory insufficiency most often have dips in SpO₂ below 95%. Symptomatic hypercapnic patients benefit from the use of noninvasive ventilation for at least part of the day, most often overnight. With progressive ventilatory muscle weakness, withdrawal of periods of daily or nightly aid for these patients will eventually re-

sult in ventilatory failure.

Neuromuscular disease patients with primarily ventilatory impairment, respiratory morbidity and mortality are a direct result of failing to assist inspiratory and expiratory muscle function as needed. For patients with primarily oxygenation impairment, the respiratory muscles, although not primarily involved, can be placed at a mechanical disadvantage by the development of lung and chest wall deformities, weakened by malnutrition and overuse, and strained to their limits by the need to ventilate stiff, noncompliant, diseased lungs or irreversibly obstructed airways. Overwork, relative or absolute, can eventually lead to secondary respiratory muscle dysfunction and overt respiratory failure. Patients with primarily ventilatory impairment who also have bulbar muscle dysfunction so severe that maximum assisted CPF are less than 160 L/m have essentially irreversible upper airway obstruction.

Ventilatory insufficiency and impaired airway secretion clearance are common

complications of spinal cord injury (SCI) and can lead to respiratory failure both at the time of the acute injury and subsequently. Respiratory complications are also a leading cause of death for these patients. Standard invasive management options such as translaryngeal intubation, tracheostomy, and electrophrenic respiration (EPR) have been used to manage these complications. In particular, once intubated and felt to not be “weanable” from ventilatory support, the patients are usually told that tracheotomy is the only option.

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 negative pressure body ventilators (NPBVs) and oscillators that create atmospheric pressure changes around the thorax and abdomen, and body ventilators and exsufflation devices that apply force directly to the body to mechanically displace respiratory muscles. Negative pressure applied to the airway during expiration or coughing assists the expiratory muscles as forced exsufflation just as positive pressure applied to the airway during inhalation (noninvasive ventilation) assists the inspiratory muscles.

Certain positive pressure ventilators or blowers have the capacity to deliver CPAP. Likewise, certain negative pressure generators or ventilators used to power NPBVs can create continuous negative expiratory pressure (CNEP). CPAP and CNEP, both first described in the 1870s,¹

act as pneumatic splints to help maintain airway and alveolar patency and to increase FRC. They do not directly assist respiratory muscle activity, are rarely indicated for patients with primarily ventilatory muscle weakness, and will not be considered examples of “noninvasive ventilation”.

The intervention goals are to maintain lung and chest wall elasticity and to promote normal lung and chest wall growth for children by the use of lung and chest wall mobilization (range of motion or ROM), to maintain normal alveolar ventilation around the clock, and to maximize cough flows. The long term goals are to avert episodes of acute respiratory failure during intercurrent chest infections, avoid hospitalizations, and prolong survival without resort to tracheotomy. All goals can be attained by evaluating, training, and equipping patients in the outpatient setting and at home.

Goal one: Maintenance of pulmonary compliance, lung growth, and chest wall mobility

As the vital capacity (VC) decreases markedly, the largest breath that one can take can only expand a small portion of the lungs. Use of incentive spirometry or deep breathing can expand the lungs no greater than the VC. Although possibly useful, manual chest wall stretching and rocking the pelvis onto the chest to decrease costovertebral tightness² has not been shown to increase lung volumes. Like limb articulations and other soft tissues, the lungs and chest wall, too, require regular ROM to prevent chest wall contractures and

lung restriction. As has been recognized since at least 1952, this can only be achieved by air stacking, by providing deep insufflations (via the upper airway or by “sighs” for patients using invasive mechanical ventilation), or by nocturnal noninvasive ventilation for patients who can not cooperate with air stacking or insufflation therapy.²

A patient’s maximum insufflation capacity (MIC) is determined by measuring spirometrically the largest volume of air that a patient can hold with a closed glottis. The patient air stacks via a mouth piece consecutively delivered volumes from a volume cycled ventilator or a manual resuscitator (Figure 1). This is performed multiple times three times daily. The patient stacks the consecutively delivered volumes with a closed glottis until the lungs are maximally expanded. If the lips or cheeks are too weak to permit air stacking, stacking is done via a nasal interface or lipseal. Patients who learn glossopharyngeal breathing (GPB) can often air stack, without mechanical assistance, consecutive GPB gulps to or beyond the MIC obtained by mechanical air stacking. This is the glossopharyngeal breathing maximum single breath capacity (GPmaxSBC). The extent to which the MIC or GPmaxSBC is greater than the VC predicts the capacity of the patient to be maintained by noninvasive rather than tracheostomy ventilatory support.³ This is because the MIC /VC difference, like the extent of assisted CPF, is a function of bulbar muscle integrity. For most patients with NMD, while the VC decreases with time the MIC increases for years before declining.⁴ If the patient’s facial



Fig. 1. Patient with neuromuscular disease performing “air stacking” with a manual resuscitator for chest wall expansion.

muscles or glottis are too weak for effective air stacking, single deep insufflations are provided via a Cough Assist™ at 40 to 70 cm H₂O three times daily.

The primary objectives in using air stacking or in providing maximum insufflations as lung and chest wall ROM are to: increase the MIC, to maximize CPF, to maintain or improve pulmonary compliance, to prevent or eliminate atelectasis, and to master noninvasive IPPV. Patients who can air stack can use noninvasive IPPV and avoid tracheotomy.

Air stacking may not only help maintain static lung compliance but it at least temporarily improves dynamic pulmonary compliance.⁵ The only studies in humans that have explored the effects of regimens of deep insufflations on static pulmonary compliance were instituted only after patients already had severe pulmonary restriction (VCs less than 50% of predicted normal), involved the use of insufflation pressures under 30 cm H₂O thus grossly inadequate to fully expand even normally

compliant lungs, and called for the use of insufflations for only minutes each day. There is no evidence that static pulmonary compliance was maintained or improved by such regimens.⁶ On the other hand, it has been shown that the MIC can be increased with the use of regimens designed to expand the lungs.^{7,8} In 278 spirometry evaluations of NMD patients old enough and able to air stack we found mean VC in the sitting position to be 1190.5 ml, supine to be 974.9 ml, while the MIC was 1820.7 ml. The higher volumes increase both unassisted and manually assisted CPF and they permit the patient to raise voice volume as desired. Lung ROM in this manner can also promote lung growth in children. Positioning of the patient to reduce pulmonary stress, avoidance of prolonged singular positioning, and using optimal trunk support when sitting have also been suggested to be beneficial in improving chest wall structure and reducing pectus.⁹ Since any patient who can air stack is also able to use noninvasive IPPV, if such a patient loses breathing tolerance during chest colds or when intubated for respiratory failure, he or she can use noninvasive IPPV or be extubated directly to continuous noninvasive IPPV whether or not having regained any breathing tolerance. This is extremely important for eliminating the need to resort to tracheostomy because such patients are extubated without being ventilator weaned. Extubation of a patient with little or no breathing tolerance who has not been trained in air stacking and noninvasive IPPV can result in panic, massive insufflation leakage, glottic closure, ventilator

dysynchrony, asphyxia, and possible 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. Thus, the first respiratory equipment that is prescribed for patients with ventilatory impairment is often a manual resuscitator. In general, because of the importance of air stacking, patients without air trapping who have diminished VCs use volume cycled ventilators rather than pressure cycled ventilators since the latter can not be used for air stacking. Use of an abdominal binder or palm pressure to the abdomen during maximal insufflations facilitates chest expansion for patients with paradoxical chest wall retraction.⁹

Just as the ongoing use of nocturnal high span bi level PAP prevents pectus and promotes lung growth and chest wall development for infants and small children with NMD, it has been suggested that the use of nocturnal nasal IPPV can decrease the rate of loss of VC following the VC plateau for patients with NMD.¹⁰ Attempts to maintain pulmonary compliance in this manner might slightly benefit VC for some populations. However, since the authors did not avoid tracheotomy for their patients, the benefits they achieved by using only nocturnal noninvasive ventilation were relatively minimal compared to those they could have achieved by using respiratory muscle aids to avoid tracheotomy .

Goal two: Continuously maintain normal alveolar ventilation by assisting inspiratory muscles as needed**The intermittent abdominal pressure ventilator (IAPV) and the chest shell ventilator**

The chest shell ventilator is cumbersome, inconvenient, and often not very effective for the user when seated. The IAPV, on the other hand, can be very effective. 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 (Exsufflation Belt™). The sac is cyclically inflated by a positive pressure ventilator. Bladder inflation moves the diaphragm upwards. 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. It requires a powerful ventilator (pump) to fill the air sac. Currently, the best portable pump on the market for this purpose appears to be the Achieva series (Tyco Puritan Bennett, Pleasanton, CA). If the patient has any inspiratory capacity or is capable of GPB, he or she can autonomously add volumes of air to those taken in mechanically. The IAPV generally augments tidal volumes by about 300 ml but volumes as high as 1200 ml can be obtained.¹² Patients with less than one hour of breathing tolerance usually prefer to use the IAPV rather than use noninvasive IPPV during daytime hours.¹¹ The IAPV is less effective in the presence of scoliosis or obesity. Recently,

a pediatric IAPV was described to have been created from two blood pressure cuffs¹² and IAPVs are being custom made in southern France.

Mouthpiece IPPV

Mouthpiece IPPV is the most important method of daytime ventilatory support for patients who need ventilatory support continuously and following extubation of patients who are unable to breathe autonomously. Most commonly, simple, flexed mouth pieces are grabbed by the patient's lips and teeth for deep insufflations as needed. Some patients keep the mouth piece between their teeth all day (Fig. 2). Most patients prefer to have the mouth piece held near the mouth. A metal clamp attached to a wheelchair can be used for this purpose or the mouth piece 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 1000 to 2000 ml. The patient grabs the mouth piece with the mouth, thereby, supplementing or substituting for inadequate autonomous breath volumes. Some patients prefer the comfort of custom orthotic mouth pieces. The patient varies the volume of air taken from ventilator cycle to ventilator cycle and breath to breath to vary tidal volume, speech volume, and cough flows, as well as to air stack to fully expand the lungs to maintain lung and chest wall compliance. To use mouthpiece IPPV effectively and conveniently, adequate neck rotation and oral motor function are necessary to grab the mouth piece and receive IPPV without insufflation leakage. To prevent the latter, the soft palate must move poste-

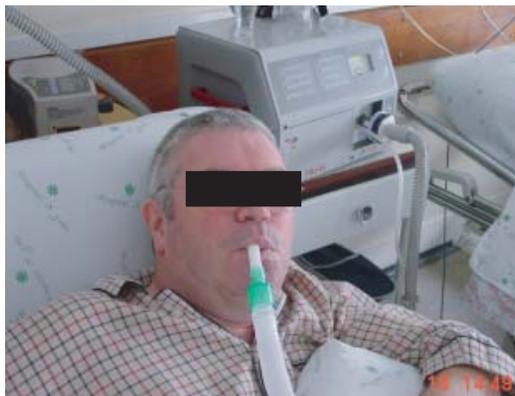


Fig. 2 – Patient with non-bulbar amyotrophic lateral sclerosis using a mouthpiece with a volume cycled ventilator for daytime ventilatory support.

riocaudally to seal off the nasopharynx. In addition, the patient must open the glottis and vocal cords, dilate the hypopharynx, and maintain airway patency to receive the air. These normally reflex movements may require a few minutes to relearn for patients who have been receiving IPPV via an indwelling tube, especially one with an inflated cuff, because reflex abduction of the hypopharynx and glottis is lost during invasive IPPV. Often patients are thought to have tracheal stenosis or other reasons for upper airway obstruction before they learn to re open the glottis to permit IPPV. Patients, and especially, children, may fail extubation to noninvasive IPPV because of upper airway obstruction due to laryngeal edema. This problem can often be avoided by glucocorticoid therapy prior to extubation. Since the low pressure alarms of volume cycled ventilators can often not be turned off, to prevent their sounding during routine daytime IPPV when not every delivered volume is received by the patient, a flexed 15 mm mouth piece for IPPV or

an in line regenerative humidifier can be used. These create 2 to 3 cm H₂O back pressure which is adequate to prevent low pressure alarm sounding.

Nasal IPPV

Nasal is preferred over lipseal ventilation by over 2 out of 3 patients who use noninvasive ventilation only for sleep.¹³ Nasal ventilation can be provided as bi level PAP or as nasal IPPV with or without PEEP using volume cycled ventilators. There are now numerous commercially available nasal interfaces (CPAP masks). Each interface design applies pressure differently to the paranasal area. One can not predict which model will be most effective and preferred by any particular patient. Nasal bridge pressure and insufflation leakage into the eyes are common complaints with several of these generic models. Such difficulties resulted in the fabrication of interfaces that mold themselves to facial tissues and in custom molded interface designs. Creative interface designs include the retention system providing the air delivery. A newly available nasal interface, called the Nasal Aire™ (InnoMed Technologies, Boca Raton, Fl), comes in 5 sizes from extra large to extra small and with its comfortable, light weight, and very practical air delivery system, has the potential to become the most popular interface on the market (Figure 3). The nose piece requires minimal pressures on the retention tubes to seal the nostrils to prevent insufflation leakage. It is also an ideal interface for patients who require noninvasive ventilation continuously because it does not interfere with the user's vision. Smaller



Fig. 3 – Patient using Nasal Aire™ (InnoMed Technologies, Boca Raton, FL).

sizes are needed, however, for infants and small children.

Since everyone's face, and especially nose, has different anatomy, one can not predict which interface will provide the best seal with least insufflation leakage, or with which interface any particular patient will be most comfortable. Therefore, no patient should be offered and expected to use only one nasal interface anymore than one should be offered only lipseal or a single oronasal interface. Alternating interfaces nightly alternates skin pressure sites, minimizes discomfort, and is to be encouraged.

Excessive insufflation leakage via the mouth is prevented by keeping ventilatory drive intact by maintaining normal daytime CO_2 and avoiding supplemental

O_2 and sedatives. However, in the presence of daytime hypercapnia and excessive nocturnal SpO_2 desaturations and bothersome arousals, for patients not wishing to switch to lipseal IPPV, a chin strap or plugged lipseal without mouth piece can be used to decrease oral leakage. In the presence of nasal congestion, patients either use decongestants to permit nasal IPPV, switch to lipseal ventilation, or on rare occasions, use a body ventilator. Most often the patient continues nasal IPPV using decongestants.

Oral nasal interfaces

Oral nasal interfaces can have strap retention systems like those for mouthpiece or nasal IPPV. Respironics Inc. and HealthDyne (Minneapolis, MN) produce comfortable strap retained oro nasal interfaces. The former manufactures a transparent total full face mask with an inner



Fig. 4 – Patient using the total full face mask (Respironics Inc).

gasket that creates a hermetic seal around the nose and mouth (Fig. 4). It was reported in one center to be more comfortable and provide a better seal than commonly available nasal interfaces.¹⁴ Custom prepared strap retained oral nasal interfaces are also available.^{15,16} These oro nasal delivery systems can be comfortable alternatives to lipseal or nasal IPPV. However, since effective ventilatory support can usually be provided by either nasal or mouthpiece/lipseal IPPV, strap retained oral nasal interfaces have been used for long term ventilatory assistance in few centers. They are being used more frequently in the intensive care setting.¹⁷

Goal three: Facilitate airway clearance by providing functional coughs by assisting expiratory muscles

Approaches to preventing peripheral airway secretion retention for patients with NMD include the use of medications to reduce mucus hypersecretion or to liquefy secretions, and facilitation of mucus mobilization. The latter can include manual or mechanical chest percussion or vibration, direct oscillation of the air column, and postural drainage. The goal is to transport mucus from the peripheral to the central airways from where it can more easily be eliminated by manual assisted coughing and mechanical assisted cough (MAC).

People with NMD and SCI, however, usually have functional bulbar musculature and normally secretory airways. They just do not have adequate inspiratory and expiratory strength for sufficient cough flows. Thus, instead of overempha-

sizing the effort intensive use of chest percussion, these patients mostly need to learn how to normalize their cough flows by using inspiratory and expiratory muscle aids. Chest physical therapy should not be used routinely for well children or anyone without excessive airway secretions. Chest clapping also causes hypoxia so at times supplemental oxygen needs to be administered.

Nasotracheal suctioning, the last resort, is considered for patients with fixed upper or lower airway obstruction and never for patients with predominantly ventilatory impairment who can use MAC effectively. To perform it, a catheter is passed via the nose or mouth into the trachea or a main stem bronchus and mucus is suctioned. It is much easier to enter the right rather than the left main stem bronchus for anatomical reasons.¹⁸ During the procedure saline may be instilled through the suction catheter into the trachea in order to liquefy the mucus. Because of triggering the cough reflex, the instillations may only reach the trachea, and may not, therefore, have any liquefying or diluting effect on mucus lower in the airway.¹⁹ The direct bronchoscopic aspiration of secretions can be preferable to endotracheal or nasotracheal suctioning because the bronchoscope permits visual inspection of the airway and direction of the suction pressure. Once airway secretions are mobilized centrally, they are ideally eliminated by MAC.

Manually assisted coughing

The importance of the use of manually assisted coughing to permit effective long term use of noninvasive ventilation is

being increasingly recognized by NMD clinic physicians and others.^{20,21} CPF are increased by manually assisted coughing. If the VC is under 1.5 L, insufflating the patient to the MIC is especially important to optimize cough flows. Once the patient takes a breath to at least 1.5 L, maximally air stacks, or is maximally insufflated, an abdominal thrust is timed to glottic opening as the patient initiates the cough. It was recognized as early as 1966 that assisted CPF could be doubled and readily exceed 6 L/s in patients receiving maximal insufflations prior to manual thrusts.²² 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 while CPF 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.

Although an optimal insufflation followed by an abdominal thrust provides the greatest increase in CPF, CPF can also be significantly increased by providing only a maximal insufflation or providing only an abdominal thrust without a preceding maximal insufflation (Fig. 5). Interestingly, CPF are increased significantly more by the maximal insufflation than by the abdominal thrust.

Manually assisted coughing requires a cooperative patient, good coordination between the patient and care giver, and adequate physical effort and often frequent application by the care giver. It is usually ineffective in the presence of severe scoliosis because of a combination of restricted lung capacity and the inability to effect diaphragm movement by abdominal thrusting because of severe rib



Fig. 5 – Spinal cord injury patient submitted to optimal insufflation with a manual resuscitator followed by an abdominal thrust performed by trained respiratory physiotherapists to optimize peak cough flow's.

cage and diaphragm deformity.

Abdominal compressions should not be used aggressively for 1 to 1.5 hours following a meal, however, chest compressions can be used to augment CPF. Chest thrusting techniques must be performed with caution in the presence of an osteoporotic rib cage.

Unfortunately, since it is not widely taught to health care professionals¹⁰⁰⁷ manually assisted coughing is under utilized.²³ When inadequate, and especially when inadequacy is due to difficulty air stacking, the most effective alternative for generating optimal CPF and clearing airway secretions is the use of MIE.

The inability to generate over 2.7 L/s or 160 L/m of assisted CPF despite having a VC or MIC greater than 1 L usually indicates fixed upper airway obstruction or severe bulbar muscle weakness and hypopharyngeal collapse during coughing attempts. Vocal cord adhesions or paraly-

sis may have resulted from a previous translaryngeal intubation or tracheostomy. Since some lesions, especially the presence of obstructing granulation tissue, can be corrected surgically, laryngoscopic examination is warranted.

Mechanical insufflation exsufflation (MI-E)

Mechanical insufflator exsufflators (Cough Assists™) deliver deep insufflations followed immediately by deep exsufflations. The insufflation and exsufflation pressures and delivery times are independently adjustable. Insufflation to exsufflation pressures of +40 to -40 cm H₂O²⁷ delivered via oronasal interface or normal adult tracheostomy tubes are usually the most effective and preferred by most patients. Lungs are insufflated until fully expanded and then immediately exsufflated until the lungs are fully emptied and the chest wall retracted. Normal cough and exsufflation volumes exceed 2 liters in adults.

Mechanical insufflation generates 2 flow notches. Onset of insufflation generates a flow peak. A second insufflation flow notch occurs when exsufflation terminates and is due to the reversal of air flow as air re enters the lung and the lung volume returns to its functional residual capacity. Mechanical exsufflation also generates 2 (exsufflation) flow notches. One occurs when the insufflation pressure stops and is due to the elastic recoil of the lung. The second one, a bit greater, is caused by the exsufflation pressure itself. Except after a meal, an abdominal thrust is applied in conjunction with the exsufflation. The combination of MI E with an abdominal

thrust is a mechanically assisted cough (MAC). Mechanical insufflation can be provided via an oral nasal mask (Fig. 6), a simple mouth piece, or via a translaryngeal or tracheostomy tube. When delivered via the latter, the cuff, when present, should be inflated.

The Cough Assist™ can be manually or automatically cycled. Manual cycling facilitates care giver patient coordination of inspiration and expiration with insufflation and exsufflation, respectively, but it requires hands to deliver an abdominal thrust, to hold the mask on the patient, and to cycle the machine²⁶.

One treatment consists of about five to six cycles of MI E or MAC followed by a short period of normal breathing or ventilator use to avoid hyperventilation. While insufflation and exsufflation pressures when used via the upper airway are almost always from +35 to +60 cm H₂O to -35 to -60 cm H₂O, it must be kept in mind that the goal is for rapid maximal chest expansion followed immediately by rapid lung emptying, both in about 1 to 3



Fig. 6 – Patient using MI-E with the CoughAssist™ via an oro-nasal mask.

seconds. The machine pressures required for this will depend on the caliber of the interface between the machine tubing and the airways. Thus, the use of MI E via narrow gauge tubes may necessitate the application of pressures to 70 cm H₂O until the endpoints are clinically observed²⁸. Most patients use 35 to 45 cm H₂O pressures for insufflations and exsufflations via the upper airway or via wide gauge adult size tracheostomy or translaryngeal tubes. In experimental models, +40 to 40 cm H₂O pressures have been shown to provide maximum forced deflation VCs and flows.²³ In a recent study in a lung model with normal human compliance, insufflation volumes exceeded 90% of predicted inspiratory capacity when insufflation times were over 5 seconds. In reality, because of lung inertial factors, pressures of 40 cm H₂O (via an interface with adequate caliber) can take a full minute to fully inflate lung tissues.²⁴

When MI E is used for airway secretion clearance, multiple treatments are given in one sitting until no further secretions are expelled and any secretion or mucus induced dSpO₂s are reversed. Use can be required as frequently as every few minutes around the clock during severe chest infections²⁶. Although no medications are usually required for effective MI E in neuromuscular ventilator users, liquefaction of sputum using heated aerosol treatments may facilitate mucus elimination when secretions are inspissated.

We routinely use MI E via oro nasal interfaces (especially in the immediate post extubation period for respiratory failure) or via translaryngeal and tracheostomy

tubes in children with SMA under 1 year of age. For infants and others who use MI E via narrow gauge pediatric tubes, even the maximum flow pressure capabilities of the Cough Assist™ are often inadequate to rapidly expand and empty the lungs. The severe pressure flow drop off across the narrow tubes greatly diminishes the efficacy of MI E for expelling airway secretions and more powerful units need to be developed. In addition to use for assisted coughing, the importance of using the Cough Assist™ for the regular provision of full chest expansion should not be underestimated.

A recent study measured flow, pressure relationships through endotracheal and tracheostomy tubes of various diameters.²⁵ Although only positive pressures were used, drop offs in pressures and flows with tube narrowing will be comparable whether using positive or negative pressures. Flows up to 200 L/m, only one third of the maximum flows emanating from the Cough Assist™, were measured. Providing 40 cm H₂O air pressure through a tracheostomy tube of 8.5 mm diameter resulted in flows of 170 L/m whereas the same pressure via a 4.5 mm tube (size of a mini tracheostomy tube, 1/2 mm smaller than a small adult tube) resulted in a flow of only about 65 L/m. The same 40 cm H₂O pressure via an 8.5 mm endotracheal tube achieved a flow of 200 L/m whereas via a 5 mm tube it resulted in only 70 L/m and via a 4.5 mm tube the resulting flow was only 55 L/m. Infant tubes are even narrower so drop offs would be even greater. In another study of pressure volume relationships through various sized tracheostomy tubes using a lung model,

alveolar pressures and ventilated volumes via 4, 4.5, and 5 mm tubes were about 40, 50, and 60% of those values via 8 mm endotracheal tubes in the absence of leakage. To obtain an insufflated volume of 500 ml, the inspiratory pressures needed were 40, 30, and 20 cm H₂O for the 4, 4.5, and 5 mm tubes, respectively.

Whether via the upper airway or via indwelling airway tubes, routine airway suctioning misses the left main stem bronchus about 90% of the time. MIE, on the other hand, provides the same exsufflation flows in both left and right airways without the discomfort or airway trauma of tracheal suctioning and it can be effective when suctioning isn't. Patients almost invariably prefer MIE to suctioning for comfort and effectiveness and they find it less tiring.²⁵ Deep suctioning, whether via airway tube or via the upper airway, can be discontinued for most NMD patients.

The oximetry feedback respiratory aid protocol

Since supplemental oxygen is avoided for NMD patients, the patient and care providers are instructed that, once artifact is ruled out,²⁹ SpO₂ below 95% is due to one of three causes: hypercapnia (hypoventilation), airway encumbrment (secretions), and if these are not managed properly, intrinsic lung disease, usually gross atelectasis or pneumonia. The oximetry, respiratory muscle aid protocol consists of using an oximeter for feedback to maintain SpO₂ greater than 94% by maintaining effective alveolar ventilation and airway secretion elimination.

The protocol is most important during

respiratory tract infections and when extubating patients with little or no breathing tolerance. Because respiratory muscles are weakened and bronchial secretions are profuse during chest infections and post surgical general anesthesia,³⁰ patients often need to use noninvasive IPPV continuously at these times both to maintain alveolar ventilation and for air stacking to increase CPF. If, when using noninvasive IPPV at adequate delivered volumes the SpO₂ is not above 94%, the desaturation is not due to hypoventilation but to airway mucus accumulation. Indeed, sudden dSpO₂s for ventilator users during chest infections are almost always due to mucus accumulation. Manually assisted coughing with air stacking as needed and MAC are then used until the mucus is expelled and SpO₂ returns to normal or to baseline levels.

If the baseline SpO₂ decreases below 95% despite ventilatory support and MAC, the outpatient then presents for a formal evaluation. A clear radiograph in the presence of a baseline SpO₂ of 92 to 94% often denotes microscopic atelectasis. Radiographic evidence of pneumonia accompanied by dSpO₂ and ventilator dependence warrants admission and, when there is respiratory distress, possible intubation. Despite hyperpyrexia and elevated white blood cell counts, baseline SpO₂ often returns to normal and hospitalization can be avoided when patients use respiratory muscle aids with oximetry feedback.³¹ As the SpO₂ baseline returns to normal, most patients gradually wean to nocturnal only noninvasive IPPV, or at times, to no daily use of respiratory muscle aids until the next chest infection.

In the event of hospitalization, the patient's primary care providers are asked to continue to use MAC with oximetry feedback to eliminate airway secretions and avoid intubation. This is done because we have found it impossible to expect nursing and respiratory therapy staff to do this as often as needed either to avoid intubation, during intubation, or post extubation. It is most practical and efficacious to let the family and care providers administer the respiratory muscle aids up to every 5 minutes if necessary to expulse secretions and return SpO₂ to normal, even when the patient is in intensive care. Overnight, provided that sedatives, narcotics, and oxygen therapy are avoided, heavy mucus accumulation will arouse the patient and he or she will request MAC. The family or primary care providers quickly learn that, since they are doing most of the work anyway, the patients can most often be best and most safely cared for at home as long as baseline SpO₂ remains greater than 94%.

Decanulation and conversion to noninvasive respiratory aids

Any patient with an indwelling tracheostomy tube who has understandable speech when the tube cuff is deflated, or when using a cuffless tube, is evaluated for decanulation. Patients without severe speech and swallowing impairment, such as patients with intact bulbar neuromuscular disease or high spinal cord injury, are usually excellent candidates. Since noninvasive ventilatory support is even successful for patients with no ability to breathe, decanulation indications depend primarily on bulbar function and, in par-

ticular, on the ability to effect glottic closure and then to maintain airway patency sufficiently for CPF to exceed 160 L/m³². Somewhat lower flows are acceptable with the tube in place because air stacking is usually impossible; the tube obstructs the upper airway; and air leakage occurs around the tube and out of the tracheostomy site during measurements of CPF. The problem of tube obstruction can be alleviated by placing a fenestrated tube to increase airflow between the upper airway and the lungs. Any cuff must be deflated and the tube capped as the patient receives IPPV via mouth piece or nasal interface and attempts to air stack.

According to our experience^{28, 32} we summarized the steps for the decanulation of patients admitted to an acute rehabilitation facility :

1. The patients were medically stabilized, supplemental oxygen therapy was discontinued, and MAC used aggressively via the tube to clear secretions.
2. The patients were placed on portable volume cycled ventilators. The cuffs were completely deflated for increasing periods, hourly, until cuff deflation could be tolerated throughout daytime hours. Partial to complete cuff deflation was then used overnight. To assure adequate alveolar ventilation the delivered insufflation volumes were increased to maintain the same airway pressures as with the cuff inflated. When necessary, the patient's tracheostomy tube diameter was changed to permit sufficient leakage for speech while maintaining adequate fit to permit effective assisted ventilation with delivered ventilator volumes generally of 1 to 2 liters. The set volumes on assist/con-

tol mode, rate 10 to 12, were titrated to maintain $p\text{CO}_2$ levels between 35 to 40 mm Hg. Tracheal integrity, tracheostomy tube width, and volitional glottic and vocal cord movements determined the amount of the delivered air to enter the lungs and the amount to “leak” up through the vocal cords with each delivered volume. The insufflation leakage was used for crescendo/decrecendo inspiratory cycle speech.

A low speech volume indicated either inadequate insufflation volume, a tracheostomy tube that was too wide, or subglottic obstruction, usually by granulation tissue. For these patients, optimal insufflation volumes for normal ventilation and more effective speech were obtained by cuff removal, by placing a narrower diameter tracheostomy tube, or by surgical ablation of the granulation tissue when indicated. The expiratory valve was occasionally capped permitting continuous speech without the expense of a Passy Muir™ valve.

3. Patients were advanced to the use of 24 hour tracheostomy IPPV with a deflated cuff. The prolonged use of partial cuff deflation was discouraged because of the tendency of the nursing staff to gradually increase the amount of air in the cuff with time. Some patients were introduced to noninvasive ventilatory support by using an iron lung or chest shell ventilator with the tracheostomy tube open. Care was taken to maintain the tracheostomy tube above the iron lung collar.

4. The patients were trained in the use of mouthpiece IPPV for daytime ventilatory support with the tracheostomy tube capped (Fig. 7). Each patient learned to

close off the nasopharynx with the soft palate to prevent nasal leakage. Temporarily pinching the nostrils helped the patient to understand why this was necessary. Patients who had had their cuffs inflated for long periods of time initially maintained their vocal cords fully adducted during attempts at mouthpiece IPPV. Each patient was unaware that he was obstructing flow. Each patient successfully relearned the reflex vocal cord abduction that permitted effective mouthpiece IPPV. Nasal IPPV was also used successfully before decanulation. EtCO_2 was evaluated by sampling end tidal air under the interface and as close to the nostrils as possible.

Once daytime mouthpiece IPPV was mastered, body ventilators were occasionally used for nocturnal support with the tracheostomy tube plugged. Body ventilator use facilitated tracheostomy site closure by relieving the stoma of the posi-



Fig. 7 – Patient training with the use of mouthpiece IPPV for daytime ventilatory support with the tracheostomy tube capped.

tive expulsive pressures that occur during IPPV and by providing ventilatory support. This prevented dyspnea and panic and facilitated the learning of noninvasive IPPV.

Mouthpiece IPPV normalized speech rhythm, provided normal daytime ventilation, and permitted “air stacking” for volitional sighing, shouting, and assisted coughing when the tube fit tightly in the tracheostomy tract. The change to a fenestrated tracheostomy tube and its capping and cuff deflation often improved the efficacy of mouthpiece IPPV as well as the effectiveness of body ventilator use. It also improved speech volume while using mouthpiece IPPV or when breathing autonomously whenever possible. The fenestration of the commercially available tubes, however, was at times malpositioned against the posterior tracheal wall where invagination of the tracheal mucosa into the fenestration rendered it ineffective and caused bleeding while potentially exacerbating granulation formation. Proper positioning of the fenestration was accomplished by ordering a custom made fenestrated tube or changing to a smaller sized tube.

5. Patients were advanced to the use of cuffless tracheostomy tubes. The great majority of patients did not require a tracheostomy diameter change to maintain optimal ventilation and speech volume, however, ventilator volumes often needed to be as much as doubled to compensate the increased leakage. Then the tube was capped or at times replaced by a tracheostomy button for up to continuous practice with noninvasive IPPV for a few days before the tracheostomy site was allowed

to close. Patients tried a variety of interfaces and chose accordingly. Once comfortable with noninvasive IPPV the tracheostomy site was allowed to close.

Patients had trials of ventilatory support by the IAPV. Several trials were often necessary to determine the optimal belt size and position and the optimal buckle or Velcro™ strap pressures. Patients with measurable VCs learned to coordinate breathing with the IAPV. With the site closed, ventilator weaning was facilitated by mouthpiece IPPV taken from a mouth piece fixed adjacent to the patient’s mouth and accessible to the patient by neck rotation (Fig. 8). The patients took fewer and fewer assisted breaths as they weaned. This technique is free of the anxiety that ventilator users feel when disconnected and weaning from tracheostomy IPPV³³. Mouthpiece IPPV is used overnight with a lipseal.

Conclusion

Mechanisms by which nasal or mouthpiece/lipseal IPPV can improve the clinical



Fig. 8 – High SCI patient using mouthpiece IPPV with the tracheostomy site closed

cal picture include: resting respiratory muscles and decreasing metabolic demand, increasing tidal volumes and relieving hypercapnia, resetting chemoreceptors, opening atelectatic areas, maintaining airway patency, improving ventilation/perfusion matching, maintaining lung and chest wall range of motion and possibly compliance, improving mucociliary clearance, and most importantly, by assisting, supporting, and substituting for inspiratory muscle function. Assisted lung ventilation and normal SpO₂ can be maintained indefinitely during sleep as well as during daytime hours for patients with little or no VC by using noninvasive IPPV

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