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Equipment Options for Cough Augmentation, Ventilation, and Noninvasive Interfaces in Neuromuscular Respiratory Management

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ABSTRACT

This is a summary of the presentation on equipment options for cough augmentation, ventilation, and noninvasive interfaces in neuromuscular respiratory management presented as part of the program on pulmonary management of pediatric patients with neuromuscular disorders at the 30th annual Carrell-Krusen Neuromuscular Symposium on February 20, 2008. *Pediatrics* 2009;123:S226–S230

THE WIDESPREAD USE of noninvasive ventilation (NIV) developed with the polio epidemics during the 1940s and 1950s when the iron lung was used to provide negative-pressure ventilation. Although negative-pressure ventilation has been an effective means of supporting neuromuscular respiratory insufficiency, its effectiveness can also be limited by neuromuscular induced upper airway instability that can limit inspiratory flow with negative pressure. Negative-pressure ventilation became a lesser option with the development of positive-pressure ventilation that would support both upper airway stability and provide adequate ventilation by noninvasive means. The first use of positive-pressure ventilation was described by Affeldt¹ in 1953, during which time patients transferred from the iron lung could be supported on intermittent positive pressure via a mouthpiece. The benefit of NIV for the nocturnal support of patients with neuromuscular disorders and respiratory insufficiency by means of both bilevel-pressure-support and volume-cycled ventilation was first described in the late 1980s.^{2–5} The application of home NIV has grown rapidly with the development of bilevel-pressure ventilation technology, noninvasive interface materials technology, and design during the past 20 years.

NONINVASIVE BILEVEL-PRESSURE VENTILATION

Noninvasive bilevel-pressure ventilation (NBPV) is based on a flow generator, sensor technology that produces 2 pressure preset levels of airflow through a single-limb ventilator circuit and into a vented nasal, oronasal, or oral interface to provide the user with a desired amount of pressure-support ventilation. The higher inspiratory positive airway pressure (IPAP) is triggered when a change in flow occurs with the patient's inspiratory effort. The flow generator cycles to the lower expiratory positive airway pressure (EPAP) when the circuit flow rate changes with the initiation of expiration. A minimum level of EPAP is necessary to flush out the patient's exhaled gas from the vented interface. The difference between bilevel pressures is the level of pressure support provided to the patient. NBPV provides a means of augmenting the ventilation of patients with respiratory insufficiency. The use of home NBPV to support patients with neuromuscular disorders and chronic respiratory insufficiency has grown rapidly over the past 10 to 15 years. Much of this growth is partly a result of technology-related improvements in bilevel-pressure generator reliability, size, and features that support the patient's use of NBPV in the home (Fig 1 www.pediatrics.org/content/vol123/Supplement_4). Integrated heated humidification with patient-adjustable temperature settings is now a standard. Most of the newer-model bilevel systems are AC and DC power compatible, and some manufacturers have battery power packs available for portability as well as emergency power supply. Perhaps the most important development in bilevel-pressure technology is in the number of adjustable settings that can be used to synchronize the bilevel-pressure ventilator to the patient's breathing pattern. Developing patient-ventilator synchrony is key to the successful application of NBPV, because this therapy is applied to patients in a conscious state. The ability to adjust the rise time (the duration of transition time between EPAP and IPAP with the initiation of an inspiratory effort) is an important factor in developing patient-ventilator synchrony. Many ventilators now have inspiratory and expiratory trigger sensitivity settings that also help the clinician to synchronize ventilator response to the patient's breathing pattern. Adjustable minimum and maximum inspiratory time settings can also be helpful in developing ventilator synchrony with the patient's breathing pattern. Although the technology improvements in bilevel-

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Abbreviations

NIV—noninvasive ventilation
 NBPV—noninvasive bilevel-pressure ventilation
 IPAP—inspiratory positive airway pressure
 EPAP—expiratory positive airway pressure
 S/T—spontaneous/timed
 VCV—volume cycled mask ventilation
 MPV—mouthpiece ventilation
 MIE—mechanical in-exsufflation
 cwp—centimeters of water pressure

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pressure ventilation have resulted in improved ventilator systems with more parameters of adjustment, it is the clinician's understanding of both neuromuscular respiratory pathophysiology and how to apply bilevel-pressure technology that is key to providing optimal respiratory support for this patient population.

Bilevel-pressure systems with a spontaneous/timed (S/T) mode setting are classified as respiratory-assist devices with an adjustable backup rate and are considered to be ventilators as compared with bilevel-pressure systems with only a spontaneous mode. The S/T mode is a necessary part of providing adequate ventilation for patients with neuromuscular disorders who develop nocturnal hypoventilation, particularly during rapid eye movement (REM) sleep. The spontaneous aspect of the S/T mode allows the patient to trigger pressure-support breaths as needed. The timed aspect of the S/T mode provides a minimum backup rate of breaths per minute when the patient is not able to trigger the ventilator at a respiratory rate above the set timed rate. Depending on the degree of neuromuscular weakness, patients with global inspiratory muscle weakness may not be able to trigger pressure-support breaths during REM sleep when the diaphragm is the only active inspiratory muscle.^{6,7} The backup rate ensures that patients will receive a minimum set number of IPAP cycles per minute when they cannot otherwise trigger the ventilator themselves.

A newer generation of bilevel-pressure servo ventilation systems have been developed to provide automatic IPAP titration in response to periodic hypoventilation patterns as found in cardiogenic Cheyne-Stokes breathing and complex sleep disordered breathing that is not supported by bilevel-pressure systems with a backup rate. Although bilevel servo ventilation has been found to be effective in maintaining a plateau of ventilation support through periodic hypoventilation patterns, it was not designed to maintain a desired level of ventilation with the onset of progressive nocturnal hypoventilation, as found in patients with chronic and progressive neuromuscular respiratory muscle weakness. Another newer generation of bilevel-pressure ventilation has been designed with an automatic IPAP titration capability based on a preset targeted tidal volume. A minimum and maximum IPAP is set along with an EPAP and S/T backup rate in conjunction with the preset target tidal volume. As the patient develops a pattern of hypoventilation during sleep, IPAP will automatically increase to maintain an average tidal volume according to the preset tidal volume. This technology holds the potential to provide patients with neuromuscular disorders who have chronic progressive respiratory weakness with a means of automatically maintaining an adequate level of nocturnal ventilation over time. Clinical studies will be necessary to determine if this technology can be effective in managing nocturnal respiratory support for patients with neuromuscular disorders and progressive respiratory insufficiency.

BILEVEL-PRESSURE INTERFACES

Commercially available bilevel-pressure interfaces are primarily nasal and oronasal designs and require an

exhalation valve either integrated in the mask shell or at the distal end of the breathing circuit immediate to the mask. It has been estimated that a 50% failure rate in compliance with noninvasive positive airway pressure therapy is attributable to difficulties related to fit and comfort with the mask interface. Fortunately there have been significant improvements in both mask designs and in the plastic materials technology in developing more comfortable and alternative mask types. Mask designs include nasal and oronasal shells, nasal pillow, nasal cannula, and oral seal masks (Fig 2 www.pediatrics.org/content/vol123/Supplement_4). There are also multiple types of mask seal interfaces, that portion of the mask that is in contact with the facial surface. These include air-cushion seal, gel interfaces, and a combination of both gel and air cushion (Fig 3 www.pediatrics.org/content/vol123/Supplement_4). Improvements in mask design have resulted in both improved comfort and decreased complications associated with mask pressure on the nasal and facial skin surfaces. Newer mask and headgear designs have also decreased claustrophobia-related discomfort with the development of nasal pillow and nasal mask shell designs that have resulted in less obstruction to the field of vision. The chin strap is also an important component in preventing oral air leak, which decreases the effectiveness of NIV when used in conjunction with a nasal mask.⁸ There are a variety of commercial chin-strap designs that can be incorporated with nasal masks to manage oral air leak (Fig 4 www.pediatrics.org/content/vol123/Supplement_4). A number of oronasal mask designs have incorporated a chin cup in the base of the mask to limit oral air leakage by preventing chin drop with relaxation of the jaw muscles during sleep (Fig 5 www.pediatrics.org/content/vol123/Supplement_4). There are now more than 50 different mask designs from at least 13 manufacturers in the United States and Europe. Although there are a wide variety of available bilevel-pressure masks from which to choose, the success in determining the most appropriate masks for a patient depends on the clinician's skill in evaluating the patient's nasal/facial shape, sleep habits, allergic rhinitis, and claustrophobia, as well as the patient's preference of mask type.⁹ Once a mask type is determined, a patient-directed desensitization protocol may be useful in helping the patient acclimate to nocturnal NIV.¹⁰ Reevaluating mask comfort and fit is also an important part of ongoing NIV management. Patients with neuromuscular disorders who must rely on the continuous support of NIV may need to alternate mask types to manage nasal and facial skin pressure-related problems. The need to use an oronasal mask for congestion related to seasonal allergic rhinitis or upper respiratory infections should also be considered. The availability of bilevel-pressure masks for pediatric patients is very limited. To date, there is only 1 air-cushion pediatric mask available in the United States that has been approved by the US Food and Drug Administration. The majority of commercially produced pediatric masks are only available outside the United States. Pediatric clinicians must rely on the available petite versions of adult masks and often must modify the headgear to provide

NIV for the pediatric patient with a neuromuscular respiratory insufficiency. The use of NIV in younger pediatric patients has also been associated with mask-pressure-induced facial malformation and skin injury.¹¹ There is a need for more pediatric air-cushion mask and nasal pillow designs to limit the effects of mask pressure in providing NIV support for younger pediatric patients. The recognized need for more pediatric mask alternatives, the growth of pediatric sleep medicine, and continued developments in mask design and technology will result in the availability of new and better NIV masks for the population of patients with neuromuscular disorders.

VOLUME-CYCLED MASK VENTILATION

Volume-cycled mask ventilation (VCMV) has been more widely used to support home mechanical ventilation in Europe,^{3,12,13} whereas in the United States, although VCMV has been used to support the home mechanical ventilation of patients with neuromuscular disorders,¹⁴ the majority of use has been largely confined to the hospital arena. VCMV requires the use of a single-limb or "J" type of volume-cycled ventilator circuit and a nonvented mask because exhaled gas is cleared through the ventilator circuit exhalation valve (Fig 6 www.pediatrics.org/content/vol123/Supplement_4). VCMV can be supported by using either a pressure- or flow-triggered ventilator. Although VCMV has been applied by using the assist/control mode, newer-generation multimode home volume-based ventilators (Fig 7 www.pediatrics.org/content/vol123/Supplement_4) can provide a number of mask-ventilation-mediated ventilator modes including volume control, pressure control, pressure support, and synchronous intermittent mandatory ventilation. There is a growing number of vented-mask manufacturers that are now producing nonvented masks for NIV. These masks are usually identified by blue- or green-colored mask shells (Fig 8 www.pediatrics.org/content/vol123/Supplement_4) as compared with vented masks, which have transparent shells. New prototypes of multimode ventilators may allow the user to switch from 1 mode of ventilation to another with preset ventilator settings by making a single setting change. This would allow the user to use 1 mode of ventilation for nocturnal support and another for portable daytime support. VCMV has been shown to be comparable to bilevel pressure in supporting patients with neuromuscular respiratory insufficiency and may be a more effective means of ventilation for the patient who either cannot tolerate higher levels of IPAP or is no longer adequately supported on bilevel-pressure ventilation. The increased application of VCMV for home ventilation in the United States will depend on the clinician's development of skills in applying this type of NIV.

MOUTHPIECE VENTILATION

The first use of mouthpiece ventilation (MPV) was described by Affeldt¹ during the 1950s when patients were supported intermittently by positive pressure via a

mouthpiece when transferred from their continuous negative-pressure ventilation by iron lung for patient care. The addition of portable daytime MPV as a complement to nocturnal mask ventilation for patients who are in need of continuous ventilatory support was first described by Bach and Saporito.^{15,16} Toussaint et al,¹⁷ in a clinical evaluation of the long-term effectiveness of MPV in a group of patients with Duchenne muscular dystrophy, showed that daytime MPV, when combined with nocturnal mask ventilation, can provide an effective means of long-term noninvasive respiratory support compared with tracheostomy ventilation. MPV is most effective when applied by using a negative-pressure-triggered volume-cycled home ventilator and a flow-restrictive mouthpiece at the end of the single-limb breathing circuit (Fig 9). Low-pressure alarming is prevented in an open-circuit system by producing enough circuit back pressure with sufficient peak inspiratory flow against the restrictive mouthpiece according to the set tidal volume (Table 1).¹⁸ The assist/control machine rate is also set at a minimum level to prevent apnea alarming. A mouthpiece circuit support arm is also necessary to position the mouthpiece immediate to the user for ease of access. The newer home volume-cycled ventilators now available are smaller, lighter, and well suited to support portable ventilation on power wheelchairs. With adequate oral muscle strength the user can trigger a ventilator breath by making a sip effort through the mouthpiece to receive breath volumes as often as needed. By taking a series of sip maneuvers and retaining the breath volumes with a closed glottis, the user can stack breaths to hyperinflate the lungs (Fig 10). (www.pediatrics.org/content/vol123/Supplement_4.) Breath-stacking maneuvers are used to prevent atelectasis and improve cough strength. Flow-triggered ventilators with a volume-control mode can be used to support MPV but are not preferred because the flow rate must be set at a high level to prevent autocycling in an open-circuit format. A higher flow-triggered rate may limit the user's ability to trigger ventilator breaths as needed. Patients with neuromuscular disorders and a reasonably good range of head motion as well as good oropharyngeal strength are likely to benefit the most from this means of portable daytime NIV.

COUGH-AUGMENTATION THERAPY

Cough augmentation is a necessary part of the noninvasive respiratory management of patients with neuromuscular disorders with respiratory insufficiency. Most patients with neuromuscular disorders do not have intrinsic lung disease that can limit the effectiveness of the mucociliary system in clearing secretions from the airways. Maintenance mucus-mobilization therapies that loosen secretions in the airways are generally not beneficial for this patient population unless there are chronic retained secretions as a result of limited mucociliary clearance. It is a weakness in the respiratory muscle groups resulting in limited cough strength that requires cough-augmentation therapy in this patient population.

MANUAL COUGH AUGMENTATION

Manual cough augmentation can be administered by supporting either hyperinflation or forced expiration alone or by combining both therapies to improve cough strength. Manual hyperinflation can be administered by using a self-inflating resuscitator bag (without oxygen reservoir) combined with a 1-way valve, a length of 22-mm corrugated ventilator tubing, and a mouthpiece (Fig 11 www.pediatrics.org/content/vol123/Supplement_4). For safety purposes, a second 1-way valve with the valve leaflet removed should be incorporated in the circuit distal to the first 1-way valve to prevent aspiration if the valve leaflet is dislodged with compression of the resuscitator bag. A nose clip may be necessary to administer hyperinflation if the patient is not able to prevent air leakage through the nasopharynx with hyperinflation maneuvers. Hyperinflation is administered by allowing the patient to inhale maximally followed by coordinated compressions of the resuscitator bag with successive inspiratory efforts, allowing the patient to maximally hyperinflate to a greater inspiratory volume than he or she can obtain independently. Patients with weak inspiratory muscle strength and adequate expiratory muscle strength may be able to significantly increase cough flows to clear secretions with manual hyperinflation therapy alone. Patients with adequate inspiratory muscle strength and weak expiratory muscle strength may benefit from an abdominal thrust maneuver to improve peak cough flows (Fig 11).¹⁹ Combining manual hyperinflation with the abdominal thrust maneuver has been shown to produce a higher peak cough flow than by using either therapy alone.²⁰ The effectiveness of cough-augmentation therapies can be assessed by measuring peak cough flow by using a simple peak flow meter. Manual hyperinflation may also be used as a maintenance therapy to maintain lung inflation by preventing atelectasis and improving chest wall compliance. A 2- or 3-times-daily regimen of 8 to 10 hyperinflation maneuvers with a 5-second breath hold at the end of each hyperinflation maneuver has been suggested as a maintenance therapy for pulmonary and chest wall compliance.

MECHANICAL IN-EXSUFFLATION

Mechanical in-exsufflation (MIE) can be used to support limited cough function by combining pressure-preset insufflation and exsufflation by means of a switch-activated reversible flow and an adjustable flow generator. A cough cycle is generated by providing a set time of insufflation based on a preset pressure to insufflate the lungs followed by an immediate change to a preset exsufflation pressure that produces a high peak expiratory flow to clear secretions. The CoughAssist device (Respironics Corp, Millersville, PA) (Fig 12 www.pediatrics.org/content/vol123/Supplement_4) has been shown to produce a higher peak cough flow when compared with combined manual CoughAssist therapies alone.²¹ MIE can be administered noninvasively by using either an air-cushion face-mask or mouthpiece circuit. The effectiveness of noninvasive MIE depends on the patient's control of

the glottis in preventing the obstruction of in-exsufflation air flows that can limit the benefit of therapy. MIE is administered by using preset in-exsufflation pressures. The range of in-exsufflation pressures for the CoughAssist device is ± 0 to 60 cm of water (cwp). In-exsufflation pressures of ± 40 cwp are considered optimal for adult patients according to manufacturer recommendations. Mean in-exsufflation pressures of ± 30 cwp with a range of insufflation of 15 to 40 cwp and exsufflation of 20 to 50 cwp have been suggested for the application of MIE for pediatric patients.²² In-exsufflation cycles can be administered either manually or by preset timed automatic-cycle mode depending on the model of CoughAssist device. Preset times are adjustable for insufflation, exsufflation, and a pause period between the end of exsufflation and the beginning of insufflation cycles. There are also 2 levels of insufflation flow from which to choose. The manufacturer's recommendation for insufflation and exsufflation cycle times for both pediatric and adult applications is based on the selected insufflation flow rate. The exsufflation pressure is usually set 5 to 10 cwp higher than insufflation to develop a high peak expiratory flow rate. Insufflation cycle time is usually 0.5 to 2.0 cwp longer than exsufflation to maximally insufflate the patient before initiating exsufflation. Many clinicians who are experienced in applying MIE with a variety of patients with neuromuscular disorders have developed their own preferences for time and pressure regimens that are felt to provide optimal therapy. The effectiveness of MIE may be limited in patients with a weak or enlarged tongue that may block exsufflation flow. Placing a modified mouthpiece within an air-cushion face mask (Fig 13 www.pediatrics.org/content/vol123/Supplement_4) is a means of preventing the tongue from limiting exsufflation flow and the effectiveness of therapy. MIE can also be applied via tracheostomy to clear secretions noninvasively. MIE can be applied directly to the tracheostomy tube for spontaneously breathing patients or through the ventilator circuit. Secretions can be removed from the circuit during exsufflation by incorporating an inline suction catheter. Place the tip of the inline suction catheter immediate to, but not into, the tracheostomy tube while applying catheter suction during exsufflation to clear secretions and maintain a clean breathing circuit (Fig 14 www.pediatrics.org/content/vol123/Supplement_4.) MIE via tracheostomy has been shown to be both more effective and more comfortable as compared with tracheal suctioning in groups of patients with spinal cord injury and amyotrophic lateral sclerosis with tracheostomy.^{23,24} The CoughAssist device can also be used to apply hyperinflation therapy. A twice-per-day treatment regimen using manual insufflation cycles of 5 to 6 seconds at ≥ 50 cwp can be applied to prevent atelectasis and improve chest wall compliance. A clinician/caregiver instructional CD-ROM available from the manufacturer can provide an understanding of the operating principal and how to apply MIE therapy.

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FIGURE 1
Bilevel-pressure ventilators with integrated heated humidifiers.



FIGURE 2
Examples of vented bilevel-pressure mask designs.



FIGURE 3
Examples of gel, air-cushion, and combined gel/air-cushion face-mask types.



FIGURE 4
Examples of chin-strap design alternatives.



FIGURE 5
Examples of oronasal mask designs that incorporate a chin cup to prevent oral air leak.



FIGURE 6
Single-limb ventilator circuits for a pressure-triggered home volume-cycled ventilator.



FIGURE 7
Examples of smaller and lighter multimode home volume-cycled ventilators with volume control, pressure control, and pressure-support capability.



FIGURE 8
Examples of nonvented masks with blue or green mask parts that differentiate them from vented masks.

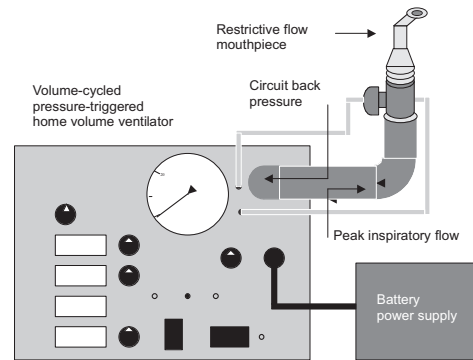


FIGURE 9
Diagram of an MPV system showing the component parts and the operating principle of flow restriction to prevent low-pressure alarming.

TABLE 1 Peak Inspiratory Flow Rate or Inspiratory Time Settings Necessary to Prevent Low-Pressure Alarming in the Respective Volume-Cycled Home Ventilators for Set Tidal Volumes of 500 to 1000 mL

Set Tidal Volume, mL	Respironics Lifecare PLV100, L/min	Mallinckrodt Achieva PSO ₂ , s	Pulmonetics LTV800, s	Newport HT-50, s	Impact Medical Univent, s	Respironics Continuum, L/min
500	55	1.2	1.7	0.8	0.8	44
550	55	1.4	1.8	0.9	0.9	44
600	55	1.6	2.0	1.0	0.9	44
650	50	1.7	2.2	1.1	1.1	44
700	50	1.8	2.3	1.1	1.1	43
750	35	1.8	2.5	1.2	1.2	43
800	35	1.9	2.7	1.3	1.3	43
850	35	2.1	2.8	1.4	1.4	43
900	35	2.1	3.0	1.5	1.5	43
950	35	2.4	3.2	1.6	1.6	42
1000	35	2.5	3.3	1.6	1.6	42

Source: Boitano LJ, Benditt JO. An evaluation of home volume ventilators that support open-circuit mouthpiece ventilation. *Respir Care*. 2005 Nov;50(11):1457-1461.



FIGURE 10 Manual hyperinflator component parts including AMBU bag, 1-way valves, 22-mm flex tubing, and mouthpiece.

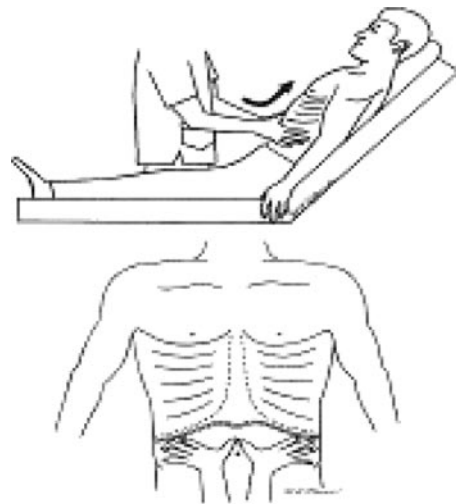


FIGURE 11 Diagram of abdominal thrust maneuver that can be used alone or in conjunction with manual hyperinflation to augment cough strength. Reprinted with permission from Braun SR et al. Improving the cough in patients with spinal cord injury. *Am J Phys Med*. 1982;63 (1): 3

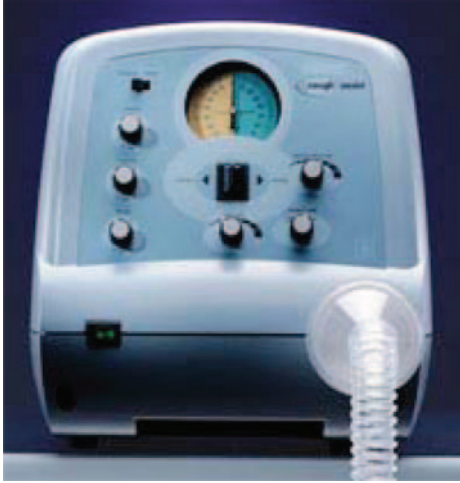


FIGURE 12
Mechanical CoughAssist device with manual and automatic modes (Respironics Corp, Murrysville, PA).

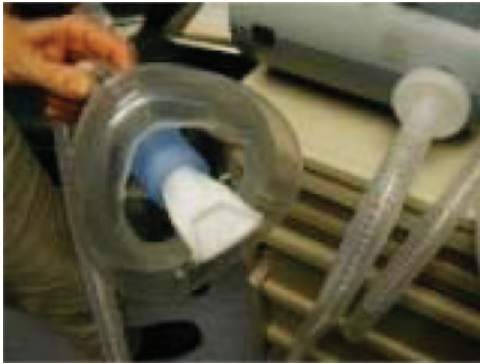


FIGURE 13
Air-cushion face mask with modified mouthpiece inserted to prevent tongue blockage with exsufflation.



FIGURE 14
Mechanical in-exsufflation applied via tracheostomy in conjunction with inline suction to remove secretions on exsufflation.

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