

FOR PATIENTS WITH INFANTILE SPINAL MUSCULAR ATROPHY (SMA) AND OTHER CONGENITAL NEUROMUSCULAR DISORDERS

(Based on the procedures developed by physicians and therapists of the Center for Ventilation Management Alternatives and Pulmonary Rehabilitation, University Hospital UMDNJ/PICU)(1-4)

These are general guidelines for the referral and subsequent management of infants with neuromuscular diseases at University Hospital/UMDNJ, Newark NJ. This protocol has significantly improved the extubation success rates of these patients over those of conventional management approaches.(1-2)

For any questions feel free to contact Brian X. Weaver RRT-NPS, RPFT at weaverbx@umdnj.edu or Dr.Bach via DoctorBach.com.

<>Candidates:

1. Children with neuromuscular disease who have developed acute respiratory failure for which they have been intubated.
2. Such patients who have failed conventional extubation in their local hospitals or who have failed to satisfy ventilator weaning criteria for conventional extubation and whose parents have decided against tracheotomy.

Referral Procedure:

1. A call must be made to the admitting resident of the pediatric intensive care unit of University Hospital at 1-973-9723784, 9723798, 9726817 or 9726827. The resident will notify the attending intensivist on call, Dr. Bach at extension .2850, and refer the patients' relatives to the Admissions Department at extension 4050. No child in distress will be accepted if not intubated. All transfers will be the responsibility of the referring institution and following management here the patient will be transferred back to the referring institution to facilitate discharge home (home nursing care, therapy, etc.).

Background Information

The Center for Ventilation Management Alternatives has developed the use of inspiratory and expiratory muscle aids that permit us to safely extubate patients who can not breathe unaided. Up to continuous use of noninvasive mouthpiece/Lipseal/nasal/oronasal ventilation is used as needed in conjunction with mechanically assisted coughing (mechanical insufflation-exsufflation with exsufflation-concomitant thoracoabdominal compression). These methods have enabled us to have a greater than 85% success rate at successfully extubating and discharging home children with SMA type 1 without tracheostomy tubes whereas the conventional extubation success rate is about 6%.(1,2)

Intubation Management Guidelines

administration is limited - only to allow the patient to approach 95% SpO₂.

This is reserved for patients with lung disease in addition to the neuromuscular issues. It may also be used for increasing SaO₂ in preparation for airway clearance. The typically routine indiscriminate use of supplemental O₂ negates the value of oximetry in gauging alveolar ventilation and airway secretion management. It can also depress ventilatory drive and, thereby, render noninvasive ventilation unsuccessful by permitting excessive insufflation leakage.(3,4)

Patients with SMA should have only ventilation, not oxygenation impairment. Secretions interfering with ventilation will reduce SpO₂. The only solution is to expel the secretions. Using O₂ to increase saturation will allow the secretions to increase and further decrease ventilation. The SaO₂ may be normal and give no indication of impending respiratory failure. Our personnel clear the airway secretions of both the right and left lungs by using in-exsufflation via the indwelling tube rather than by only suctioning via the tube, an intervention that almost invariably results in clearing the right airways but not the left airways.

Respiratory Therapy Interventions

Mobilization of secretions:

Place the patient in a postural drainage position.

The position and angle should be appropriate for the lobes and degree of severity of secretions and or plugs.

Postural drainage should be used for all lung secretions that require aggressive clearance methods. It is usually required in the morning and at night. Maintenance therapy does not always require postural drainage.

Loosening secretions:

CPT must be aggressive and should follow the AARC guidelines. The approach should be similar to that applied to a cystic fibrosis patient.

12-point CPT should be performed with 1 to 3 minutes of percussion applied to each lobe. The amount of time depends on the amount of secretions.

3 min for morning and periods of plugging and congestion.

Maintenance usually only requires 1 minute or 2 minutes.

Mechanical insufflation-exsufflation (the Cough-Assist™) is used via the tube, if the patient is intubated, or via a resuscitation mask, if non-invasively ventilated, at about +40 cm H₂O to -40 cm H₂O pressures. The goal is for rapid (2-4 sec) expansion to rapid emptying of the lungs.

Each session of airway clearance consists of several sets of multiple cycles of insufflation-exsufflation. Sets of 3, for basic maintenance, may be increased to 5 for aggressive secretion expulsion.

Three cycles are consecutively applied without removing the mask for suctioning for routine maintenance, and increased to 5 when you need to be more aggressive.

Insufflation and exsufflation are usually at the same pressure and duration. The duration consistency is achieved by using a cadence of "1-2-3" spoken out loud followed by the word "COUGH" and finally followed by "1-2-3". This is an important part of the process because the patient and CoughAssist™ therapist must work in unison.

Abdominal thrusts are applied during the exsufflation phase of each cycle.

The thrust is a type of Heimlich maneuver that uses the diaphragm to help compress the air in the lungs and airways to maintain airway patency and to augment the expiratory flow.

Do not push down on the belly - the motion should be upward around the belly button.

<!--[if !supportLists]-->5. <!--[endif]-->Tube and upper airway are suctioned following the use of expiratory aids (insufflation /exsufflation). Suction is required only to the back of the pharynx. Nasal suctioning should be performed after step three with a soft red catheter to reduce nasal trauma.

Additional Points

1. The first through the fifth steps are performed as needed. Scheduling routine treatments should be avoided because it may cause treatments to be done without regard for patient need or benefit. The patient's specific clearance needs determine the treatment times and intensity. Generally, the morning requires a very aggressive treatment and the evening requires a moderate treatment for sleep. Treatments in-between are indicated by vital signs and patient demand. The orders are best written as PRN intervals. Relatives and all ICU staff are an important part of the overall treatment and monitoring system. The frequency could be as little as twice a day or as much as every 10 minutes to reverse desaturations due to airway mucus accumulation.
2. One may experience a nonproductive session regardless of how hard one tries. If the vital signs are good and the patient is stable, it is best to leave the patient for a short time (maybe ½ hour). After a short time, re-attempt step three, and you will usually have very good results. This is common and most likely to occur in the morning.
3. Additional sessions may be done between normal airway clearance sessions.
4. Small amounts of secretions that develop after the session can be cleared quickly by suctioning the mouth and throat. In-exsufflation can be used with just a quick 1 or 2 cycles to get small amounts of mucus. If all signs are clear with this little effort, no further treatment will be necessary and the patient's normal activities (rest, sleep, play, etc...) may be resumed.
5. If there are signs of impending distress, then more complete and aggressive measures will be required.

Issues Related Specifically to the Intubated Patient

1. Ventilator weaning is attempted without permitting hypercapnia.
2. For a successful extubation the following pre-extubation criteria must be met:

Afebrile

No supplemental oxygen requirement to maintain SpO₂ >94%

Chest radiograph abnormalities cleared or clearing

All respiratory depressants discontinued

Airway suctioning required less than 1-2x/eight hours

Coryza diminished sufficiently so that suctioning of the nasal orifices is required less than once every 6 hours. This is important to facilitate use of nasal prongs/mask for post-extubation nasal ventilation.

3. Extubation to continuous nasal BiPAP ventilation and little or no supplemental oxygen.

Oximetry feedback is used to guide the use of expiratory aids, postural drainage, and chest physical therapy to reverse any desaturations due to airway mucus accumulation.

5. CO₂ retention or ventilator synchronization difficulties may be eliminated by improved nasal interface, increase in pressure support and ventilator rates or a change from BiPAP-ST™ to using a volume-cycled ventilator. Persistent desaturation despite eucapnia and aggressive use of expiratory aids indicate impending respiratory failure and need to re-intubate.

6. Following re-intubation, the protocol is used for a second trial of extubation to nasal ventilation. Following successful extubation, the patient may wean to nocturnal nasal ventilation.

7. Discharge home is considered after the SpO₂ remain within normal limits for 2 days and when assisted coughing is needed less than 4 times per day. Family training must begin at admission. They must be continuously trained and then be a part of the ongoing care. This is essential for the transition of the patient from the ICU to home



1. Bach JR, Niranjana V, Weaver B. Spinal muscular atrophy type 1: a noninvasive respiratory management approach. *Chest* 2000;117:1100-1105.

2. Bach JR, Baird JS, Plosky D, Nevado J, Weaver B. Spinal muscular atrophy type 1: management and outcomes. *Pediatr Pulmonol* 2002;34:16-22.

3. Bach JR (ed). Noninvasive Mechanical Ventilation. Philadelphia: Hanley & Belfus 2002, 348 pages.

4. Bach JR. The Management of Patients With Neuromuscular Disease. Philadelphia: Hanley & Belfus 2004, 414 pages.