



Treating Neuromuscular Patients Who Use Home Ventilation: Critical Issues

This patient, _____, has a neuromuscular condition that requires invasive noninvasive home ventilation. **Take note of these nine issues critical for treatment.**

1. The patient and designated caregiver are experts.

Long-term ventilatory support at home requires different skills and knowledge than acute ventilation in an emergency or intensive care unit. Long-term ventilator users frequently have years of experience in dealing with ventilatory issues and respiratory care problems and have learned what works and what does not. They know their bodies and their equipment.

It is not possible for medical providers in acute care settings to know all of the strategies patients have developed to deal with their unique problems. So it is critical for medical providers to approach these patients with an open mind and accept the patient's suggestions *even if they run contrary to standard hospital protocols*. **It is recommended that the attending physician state in the orders that the caregiver should remain at all times to participate in care.**

2. Communication is critical to successful treatment.

So if patient is conscious, please:

- **Listen.** Read lips/ provide paper and pen/ utilize patient's communication device if needed.
- **Repeat back** what you hear, providing an opportunity for the patient to clarify.
- **Talk directly to the patient.** A patient unable to communicate may still comprehend. Explain your findings and treatment plan.

3. For patients with a pre-existing tracheostomy (the interface for hyperinflation maneuvers and ventilation).

The patient's routine may include less common details such as cuffless ventilation or ventilation with a Passy-Muir™ valve in place. Additionally, serious acute illness may require changes such as temporary use of a different type of trach tube. **Please discuss with patient and caregiver and return to the patient's own routine as soon as possible.**

4. *Caution!* Providing oxygen alone may have dire consequences!

Oxygen used alone may mask or accelerate acute respiratory failure in neuromuscular patients. The response to low oxygen levels must be to increase ventilatory support and secretion management, NOT simply to administer oxygen.

continued

Give oxygen to neuromuscular patients ONLY if all four of these conditions exist:

1. There is an additional pulmonary condition such as pneumonia, COPD or pulmonary embolism, **and**
2. O₂ saturation is below 90% **and**
3. Secretion management, i.e., CoughAssist® or air stacking, has failed to improve saturation levels **and**
4. Mechanical ventilation is securely in place.

Then provide only low levels of oxygen and monitor CO₂ levels. Oximetry and EtCO₂ (End Tidal) are preferable and adequate for measurement.

Ventilation is critical.

Patients with neuromuscular disorders require VENTILATORY support. Unless the neuromuscular patient has a separate pulmonary disease, hypoxemia signals either accumulation of secretions or inadequate ventilation. START VENTILATION AND A SECRETION MANAGEMENT PROGRAM.

- Ventilation can be noninvasive via a mask or mouthpiece or provided by an existing tracheostomy with a machine-set respiratory rate. Intubation may be required temporarily in emergency situations if noninvasive ventilation is not currently effective.
- Secretion management should primarily be hyperinflation using the patient's customary method, i.e., manually assisted cough (abdominal thrust), air stacking with resuscitator bag or volume vent, or CoughAssist®. (See below.) If secretions are copious and caregivers or family are not available to use the CoughAssist®, suctioning and/or intubation may be required until patient improves enough to transition back to noninvasive ventilation.
- CoughAssist® is a portable device that many home mechanical ventilation patients, trached or otherwise, utilize to bring up secretions because they have a weak cough. This device must be by the patient's bedside and used as frequently as needed, possibly several times an hour until secretions are controlled. The patient and caregiver know how to operate this device and may prefer to operate it themselves.

5. **Caution!** Anesthesia and sedation for neuromuscular patients can dangerously decrease respiration.

Neuromuscular patients with respiratory muscle weakness develop respiratory failure due to hypoventilation. Anything that depresses their respiratory drive, such as opiates and benzodiazepines, will worsen hypoventilation and worsen respiratory failure. In noninvasively ventilated patients, sedating medications can blunt the patient's response to interface air leaks.

Ventilation may eliminate the need for sedation.

Medications such as narcotics and benzodiazepines are frequently used when patients appear to be dyspneic, anxious, uncomfortable, or "fighting the ventilator." While these medications are important and should not be withheld if needed, healthcare providers need to be aware of the patient's respiratory status. For patients who typically use noninvasive ventilation, ***dyspnea and anxiety can frequently be relieved by starting noninvasive ventilatory support.***

If anesthesia is a must ...

- It should only be administered by very experienced personnel under close monitoring.
- In the recovery room the patient should be returned to his/her home ventilation system (noninvasive or invasive) with his/her usual settings and with airway carefully monitored.

6. Using the patient's own ventilator is optimum.

The home ventilator is programmed to the settings used at home and the caregiver knows how to use it.

- *During short (less than one hour) ambulance transport, use patient's home ventilator rather than bagging. For longer trips, connect the ventilator directly to the ambulance's AC power.*
- *In hospital, use patient's home ventilator. Otherwise, use hospital's equivalent device unless a more sophisticated form of ventilation is required. (If patient's equipment is pre-approved by hospital, patient will provide documentation.)*
- Direct hospital RCP (Respiratory Care Practitioner) to consult by phone with patient's home RCP. See [Patient's Vital Information for Medical Staff](#) for contact information.

7. Not all body positions are tolerated by neuromuscular patients.

These patients often cannot tolerate positions normally assumed during treatment, such as lying on their backs. Ask the patient or caregiver about acceptable positions and ventilation needs during treatment.

8. *Important!* A tracheostomy is often unnecessary for patients previously using noninvasive ventilation.

As long as secretions can be managed noninvasively, there may be no need for tracheostomy support. Properly used noninvasive ventilation for a cooperative patient may provide as much respiratory support as invasive ventilation.

Generally, persons who are speaking and swallowing reasonably well do not need a tracheostomy for ventilation. They may require intubation temporarily for severe pneumonia or, like anyone else, for recovery from the trauma of surgery; **but they can usually transition back to noninvasive ventilation.** If possible, consult with the patient's physician. See [Patient's Vital Information for Medical Staff](#) before proceeding.

9. Life continuation/cessation is the patient's decision

This patient has completed and discussed with his/her caregiver a Living Will and Medical Power of Attorney. The health professional's responsibility in this regard is specific and limited.

The health professional's responsibility **is to provide information** that will help the patient or the person designated as Power of Attorney to make a decision. *For example: "Some tracheotomy patients can return to noninvasive ventilation over time."*

The health professional's responsibility **is not to make judgments** about the patient's situation. *For example: "Your care in this condition will be too much for your family."*

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