

Pulmonary Support for Myotonic Dystrophy Patients During COVID-19 Pandemic

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BACKGROUND

1. Myotonic Dystrophy (DM) can affect breathing function

- a. DM does not in general damage lung tissue
 - Even DM patients with weak muscles do not need supplemental oxygen when otherwise healthy
- b. Impaired breathing in DM is due to weak diaphragm and other breathing muscles
 - Breathing is supported with advanced positive airway pressure (PAP) devices including home ventilators, commonly known as noninvasive ventilation (NIV), via a mask interface. These are often used at night. During the daytime, some individuals may receive ventilation via a mouthpiece (sip ventilation).
 - Tracheostomy and a home ventilator are used when breathing muscles are very weak

2. COVID-19 can affect breathing function in all individuals, even if their muscles are strong

- a. COVID-19 does not directly affect breathing muscles, but inflames lung tissue
 - Oxygen has greater difficulty moving through the lung in COVID-19
 - COVID-19 patients with low oxygen benefit from supplemental oxygen



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Myotonic's mission is to enhance the quality of life of people living with myotonic dystrophy and accelerate research focused on treatments and a cure.

CONCERNS RAISED BY COVID-19 REGARDING BREATHING SUPPORT IN MYOTONIC DYSTROPHY

1. **DM patients with COVID-19 will require both PAP Ventilation (NIV or Intubation) AND supplemental oxygen**
2. **NIV support in COVID-19 can increase spread of viral particles to surroundings and infect others**
 - a. COVID-19 is mainly spread through droplets produced by coughing or sneezing
 - b. NIV and airway clearance devices (cough assist, nebulizer) can “aerosolize” COVID-19 virus – spreading it much more widely
 - c. To reduce viral spread, most hospitals are discontinuing routine use of NIV and airway clearance devices
 - d. Home mask interfaces are vented and can blow virus out of the CO₂ exhalation ports, spreading the virus in the surrounding environment
 - e. Masks with a high leak also increase viral dispersion
3. **Modifying NIV devices is recommended for DM patients suspected of having, or being infected by COVID-19**
 - a. Unvented well-fitted full-face masks need to be used with a dual lumen hose with a compatible ventilator
 - Without venting, CO₂ can dangerously build up in the lungs
 - A dual lumen hose allows CO₂ to be removed and limits spread of the virus
 - b. New home ventilators (e.g. Philips EVO, ResMed Astral 150, VOCSN) have dual lumen capability

BREATHING SUPPORT OPTIONS FOR MYOTONIC DYSTROPHY PATIENTS DURING COVID-19 PANDEMIC

1. At home

- a. If there has been no exposure to COVID-19 usual breathing supports are appropriate
 - Caregivers need to follow CDC guidelines closely
 - Wash hands, use $\geq 60\%$ alcohol-based sanitizers, do not touch face, avoid contact with anyone possibly infected
- b. If there is evidence of COVID-19 exposure or infection but breathing has not changed
 - Increase protection of caregivers to reduce the risk of their being infected
 - Follow cleaning recommendations for equipment closely (also see ACCP guidelines)
 - Monitor oxygenation carefully, use home pulse oximeter if possible
 - Maintain close contact with medical providers

2. Emergency department or hospital pulmonary care, if there is suspicion of COVID-19

- a. DM patients with COVID-19 infection will require PAP support (NIV or intubation) and supplemental oxygen
- b. To decrease spread of COVID-19 in the hospital, use of home PAP/NIV will likely not be allowed
- c. To avoid intubation, a double-lumen compatible ventilator can be used with an unvented, well-fitted full-face mask to provide NIV
- d. Severe pneumonia may necessitate intubation and ventilator support
- e. As hospital ventilators become scarce, specific home ventilators (e.g. Trilogy EVO, Astral 150, VOCSN) may be used, if hospital policy permits

For more details see *American College of Chest Physicians (ACCP) Care Recommendations for the Home-Based Ventilation Patient Undergoing Therapy for Known or Suspected Respiratory Viral Infection with COVID-19*

<https://www.chestnet.org/Guidelines-and-Resources/Resources/CHEST-Novel-Coronavirus-Resources>

<https://foundation.chestnet.org/patient-education-resources/>

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CHEST Home-Based Mechanical Ventilation and Neuromuscular Disease NetWork

Care Recommendations for the Home-Based Ventilation Patient Undergoing Therapy for Known or Suspected Respiratory Viral Infection with COVID-19 ^{1,2,3}

The American College of Chest Physicians (ACCP) pulmonary and lung health experts, along with the Home Mechanical Ventilation and Neuromuscular Disease NetWork would like to provide recommendations with specific guidelines addressing noninvasive positive pressure ventilation (NIPPV) and mechanical airway clearance in neuromuscular medicine patients (and those who are on home ventilation for other chronic respiratory failure syndromes), who are undergoing evaluation and therapy for known or suspected infection.

Noninvasive positive pressure ventilation and mechanical airway clearance can increase risk of dispersion of aerosolized virus into the environment. This risk is especially of concern with poor fitting mask interfaces, high leak, and open ventilation systems with tracheostomy.

- 1) **NIPPV may spread infectious particles – considerations to limit spread**
 - a. The use of a full-face mask (covering nose and mouth).
 - b. Add an additional combined bacterial/viral filter (hepa) between the mask and device tubing to reduce particle spread (add swivel CO₂ exhalation port on tubing and switch to non-vented mask).
 - i. With this option, device humidifier needs to be off
 - c. Or, place a mask over the CO₂ exhalation port of your mask (if you are using a *vented mask*) – to reduce particle spread. Caution will need to be exercised to ensure the mask does not stick to the exhalation port of the mask and occlude it, as this would cause CO₂ retention.
 - d. Or, change tubing to a closed system with a double lumen tube and non-vented full-face mask for compatible home ventilators.
 - e. Need to discuss with your DME provider for set up of above options.
- 2) **Care for disposables for ventilation devices:**
 - a. Mask interface– The leak from the mask is a significant source of infection. Daily cleaning with a cleaning wipe should be considered. (Mask cushions made from foam cannot be exposed to water and should not be used when sick.) The wipes can easily be made at home as below:

To Make Disposable Cleaning Wipes, You Will Need:

- 1 sealable container large enough to fit a roll of paper-towels cut in half
- 1 roll of the THICK paper towels. (Take the center tube out.)
- 2 cups of water, boiled and cooled
- 2 tablespoons concentrated dish soap
- 2 tablespoon white vinegar

Put towels in container and saturate with the solution. Keep container sealed.

- b. Hoses – clean the hoses with sterilizing solution every other day. This could be done with commercial solutions (such as Control III Disinfectant - <http://www.controlthree.com/>). You can also use a 50% hydrogen peroxide solution.
 - c. Humidity chambers – Place fresh water into the chamber daily (distilled/bottled or boiled). Clean the chamber and the hose every other day.
 - d. Also see ResMed and Philips cleaning guides
 - a. <https://www.resmed.com/en-us/sleep-apnea/cpap-parts-support/cleaning-cpap-equipment/>
 - b. <https://www.usa.philips.com/c-e/hs/better-sleep-breathing-blog/better-sleep/keeping-it-clean-cpap.html>
 - e. Filters –
 - **Consider adding an additional in-line combined bacterial/viral filter with the device.** See #1 above. (Filters can be obtained from online sources but are likely available from your DME provider). You should change the filter every three days while you are sick. The filter can help to reduce droplets and spread of infection to caregivers.
 - The standard device filter should also be changed once a week while you are sick.
- 3) **Care for the ventilation device**
- a. Cleaning should be done by your DME provider when you are better.
- 4) **Oral and nasal suction**
- a. Cleaning of the suction device—clean your suction canister daily with a commercial sterilizing solution or a 50% hydrogen peroxide solution.
 - b. Cleaning and changes of tubing and yankour
 - Yankour – clean daily with a commercial sterilizing solution or a 50% hydrogen peroxide solution. Between oral suctioning, consider wiping down with a paper towel or gauze with chlorhexidine solution. (EX: Peridex <https://www.3m.com/>) Get a new yankour at the end of the illness.
 - Suction catheter -- clean after suctioning with a 50% hydrogen peroxide solution. Use one catheter a day and then discard.
 - c. Consider adding additional suction adapters
 - Nose – Consider adding a small silicone adapter for nasal suction (EX: [The little sucker](https://www.neotechproducts.com/product/little-sucker/) from neotech <https://www.neotechproducts.com/product/little-sucker/>). Clean after suctioning with a 50% hydrogen peroxide solution.
 - Deep pharyngeal suctioning – Consider using a directional aid like the [No BiteV](http://www.njrmedical.com/no_bite_v.php) (available online http://www.njrmedical.com/no_bite_v.php). Clean between uses with a 50% hydrogen peroxide solution.
- 5) **How to prevent secondary infections (e.g. pneumonia):**
- a. Keep the head of your bed elevated to ~35 degree
 - b. Keep oral care aggressive including chlorhexidine solution
 - c. Consider in-line suction and once weekly tracheostomy tube changes for *invasive* mechanically ventilated patients

6) **What your family and caregivers need to know:**

- a. Space needed for isolation while using NIPPV – NIPPV is known to spread infectious particles, especially with poorly fitted masks. Well fitted full face masks covering nose and mouth limit spread (as compared to nasal or pillow mask interfaces) but may necessitate additional monitoring by caregivers in young children and/or those who cannot remove the mask by themselves. Well-fitting masks are best for limiting spread. Assuming the most conservative plan a space of one yard (3 feet).
- b. What caregivers need to do to keep themselves safe –
 - Gloves - when the caregiver is in the room of a patient, they should wear gloves - and change gloves each time.
 - Masks – when in the patient’s room – the caregiver should wear a mask (preferably N95) and eye goggle for protection, leave equipment in one location right outside of patient room, and dispose daily.
 - Caregiver should use protective gown/clothing when in room, same protocol as above.
 - Caregivers should clean surfaces with commonly available anti-bacterial/viral spray.
- c. What resources should you keep at home –
 - Extra distilled water
 - One-month extra supply of medications
 - Extra laundry supplies

7) **What should I do if I have a tracheostomy:**

- a. Tracheostomy tube change / cleaning frequency
 - Change – once a week tracheostomy tube changes while sick
 - Tracheostomy cleaning protocol –
 - Get one new tracheostomy tube every 3 months and always have at least 2 tracheostomy tubes at home.
 - Change the tracheostomy tube every week.
 - Use tracheostomy cleaning kits to clean the old tube and when it is very dry - then store it in an airtight container until it is needed for the next tube change.
 - At the end of three months the oldest tracheostomy is pitched a new tube is put in the rotation.
 - Cleaning Instructions: Clean the tracheostomy tube using the brush in the cleaning kit - put hot soapy water in the base of the kit and when brushed clean -- then rinse through with boiling water. When the tube is dry put in an airtight container and save for the next tracheostomy change in one month. (50% hydrogen peroxide solution can be used in lieu of soapy water if desired)
- b. Tracheostomy ties and stomal care— Continue with daily tie changes and change stomal dressing as needed to keep gauze and stoma dry. Remember that these are respiratory secretions and will be densely filled with infectious particles. The ties and dressings should be thrown away into a zip lock bag and sealed before putting in the trash.

- c. Consider switching to an in-line suction system – this will reduce exposure to caregivers (one less task for the caregivers) and reduce the development of secondary infections.
- d. Leak Speech Ventilation - controlling particle spread (balloon up ventilation) – Leak speech ventilation is common for those on home based **invasive** mechanical ventilation. The concern is that the high leak associated with this mode of ventilation (balloon down) significantly spreads infectious particles. Work with your physician and respiratory therapist to develop a safe alternative setting that will allow you to put in a cuffed tracheostomy and put the balloon up until you are better. If you choose this mode, you will need to plan for alternative mode of communication.

8) **If you need to go to the hospital**

- a. Avoid hospital, if possible, unless you have fevers (> 100 F) and/or increasing shortness of breath that does not respond to your usual treatment. Please also contact your pulmonologist. You should be aware that if you are admitted to the hospital, you may not be able to use noninvasive ventilation.
- b. Home ventilation devices – Bring ALL of your home devices as the hospital may not have what you are used to – and they may be out of devices. Some hospitals will not allow use of home equipment, but it is a safe precaution to bring them.
 - Know your settings (ask your provider to give you a one-page list that includes your PAP device/home ventilator settings, cough assist, suction, nebulizer therapy.)
 - **Confer with hospital medical providers to convert the device tubing/mask circuitry into a closed system (with a compatible ventilator), which is a double lumen tube with a non-vented full-face mask.** This will limit risk of infectious particle spread. (New home ventilators are capable of double lumen tubing, e.g. Philips Evo, ResMed Astral, VOCSN)
 - Know your medication regimen
 - Know your airway clearance regimen
 - Go to the “take charge not chances” program from the International Ventilator Users Network (IVUN) and fill out the following:
 - The Take Charge, Not Chances Portfolio
 - (<http://www.ventusers.org/vume/index.html>)
 - Home Ventilator User's Emergency Preparation Checklist (pdf)
 - Caregiver's Emergency Preparation Checklist (pdf)
 - Patient's Vital Information for Medical Staff (pdf) (Word option)
 - Treating Neuromuscular Patients Who Use Home Mechanical Ventilation: Critical Issues (pdf)
- c. Advocate for frequent and scheduled airway clearance - Bring your home devices (cough assist, therapy vest, etc.) and you may need to have your caregivers give you the airway clearance treatments. The hospital may only have basic suction available.
- d. Challenges around the use of oxygen – If you have neuromuscular disease, the use of supplemental oxygen can be risky, causing steep escalation in blood carbon dioxide (CO₂) levels. You may have been instructed that you should never be treated with oxygen. You should be aware that in the setting of infectious pneumonia – you may need oxygen in order to maintain adequate oxygen saturation with noninvasive ventilation. As long as oxygen is

delivered through your Positive Airway Pressure (PAP) Device or Ventilator – you will be protected as the CO₂ will be washed out by your PAP device/ventilator.

- e. Neuromuscular patients infected with COVID19 will need both NIPPV and oxygen.
- f. For patients who are severely ill or showing signs of deterioration, intubation and mechanical ventilation may be required.

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