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

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 CO_2 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, CO_2 can dangerously build up in the lungs
      - A dual lumen hose allows CO_2 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

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

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 ≥ 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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