

Guidance on Airway Clearance with Suspected SARS-CoV-2 (COVID-19) in Congenital Muscular Dystrophy-Affected Patients

Rationale:

There have been a great deal of public health policies implemented to limit transmission of SARS-CoV-2 (COVID-19). Beyond the level of social distancing is a further mitigation of risk by limiting procedures that can increase the likelihood of generating infectious airborne droplets, referred to as **Aerosol Generating Procedures (AGP)**. These include two procedures that are critical to maintaining health in affected individuals with Congenital Muscular Dystrophy (CMD):

1. Assisted airway clearance with cough assist
2. Non-invasive nasal ventilation

Standard Management of an Acute Respiratory Illness in Neuromuscular Disease:

1. During an acute respiratory illness, individuals who have a neuromuscular condition often are unable to cough and clear secretions, and need increased assisted airway clearance with use of the Cough Assist®.
2. Increased airway clearance is critically important during the early stage of a respiratory illness
3. During an acute respiratory illness there is a need for use of continued or increased non-invasive ventilation (NIV)

Aerosol Generating Procedures (AGP) Mitigation:

1. **Many hospitals seek to limit the use of AGP, such as Cough Assist and NIV in acutely ill patients**
2. This applies until the patient is COVID-19 negative.
3. Once the patient is COVID-19 negative then Cough Assist and NIV can be used at the discretion of the treating clinician
4. If the patient is COVID-19 positive, then the patient is placed in high level isolation

The Problem:

If an acutely ill CMD patient does not receive aggressive airway clearance early in the course of an acute illness there is an increased risk of:

1. Prolonged duration of the respiratory illness
2. More severe respiratory illness and death

The Solution:

1. Upon presentation to urgent care / emergency department with symptoms suggestive of COVID-19, a CMD patient should be placed in high level isolation as per institutional policy
2. COVID-19 testing should be performed
3. While COVID-19 testing is pending, the patient should be treated aggressively with airway clearance using Cough assist and NIV at the discretion of the treating physician
4. **It is not an option to limit airway clearance or NIV pending COVID-19 testing**
5. The compromise is to place the patient into isolation pending COVID-19 testing and provide appropriate airway clearance and NIV until COVID-19 testing is completed.

References:

- [CMD Pulmonary Guide for affected individuals, families and clinicians](#)
- [World Muscle Society position and advice: Covid-19 and people with neuromuscular disorders](#)
- [Chest Foundation: COVID-19 Resources: Care Recommendations for Home-Based Ventilation Patients](#)
- [Overview: Breathing in CMD](#)

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