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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