Respiratory management for patients with neuromuscular disorders during the COVID-19 pandemic

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To the Editor,

The coronavirus-19 (COVID-19) pandemic has forced a rapid and unprecedented reconstitution of care delivery worldwide. As respiratory compromises cause the majority of mortalities and morbidities of patients with rare neuromuscular disorders (NMD), emerging data suggest an accumulation of risk of exacerbation caused by COVID-19 in this specific patient group. Patients with motor neuron disease, amyotrophic lateral sclerosis, spinal muscular atrophy and various muscular dystrophies, and metabolic myopathies (e.g. Duchenne muscular dystrophy, Pompe disease), who have ventilatory muscle weakness or cardiomyopathy are likely to be at increased risk of contracting a severe COVID-19 infection.[1] Conclusively, the COVID-19 pandemic has the potential to disproportionately and severely affect patients with NMD.

Recently, several neurological associations and neuromuscular networks have produced guidance for caring for the impact of COVID-19 on patients with NMD, especially in the management of devastating respiratory compromises. Close monitoring of moderate- to high-risk patients with NMD for a possible more rapid decline in respiratory function or for a worsening of their underlying NMD is recommended. According to the suggestion of the World Muscle Society, pulmonary features conferring a high risk of severe COVID-19 in patients with NMD include: (a) a <60% predicted forced vital capacity, especially in those associated with kyphoscoliosis; (b) supported with a ventilator *via* a mask or tracheotomy; (c) a weak cough and weak airway clearance due to oropharyngeal weakness; (d) the presence of a tracheostoma. Decisions on patient admission to the intensive care unit (ICU) may be affected by anticipated or existing capacity problems, therefore, these guidelines help healthcare providers to identify the risk of a severe course of COVID-19 as high or moderately high in all but the mildest forms of NMD.[2]

At this time, although there are a few neuromuscular-specific recommendations for patients with NMD who contract COVID-19, additional outcome data are still needed. In particular, modifications of the respiratory care method for patients with NMD during the COVID-19 pandemic are urgently needed. With the tendency of a high rate of mechanical ventilator dependence, recent studies have underscored the utility of noninvasive ventilation (NIV) as a first-line intervention of acute respiratory failure in patients with NMD to avert intubation, to facilitate extubation, to shorten ICU stays, and to improve survival.[3,4] However, recent studies have restricted NIV use in the scenario of severe COVID-19 pneumonia.[5,6] Conventional mechanical ventilators are usually equipped with separate hoses for inhalation and exhalation, which are merged in a y-piece near the patient. It permits the collection of exhalations, which are then released into the room through a heat and moisture exchanger filter. However, devices for NIV usually are designed as a single circuit with only one hose. Therefore, exhalations are released without filtering *via* a valve that works like a desired leak. As a consequence, high-flow NIV might lead to a much higher aerosolized COVID-19 viral contamination than conventional ventilators. Appropriate measures with modifications might help to prevent the spread of infectious particles into the healthcare environment. Some experts suggest

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Figure 1. The innovative design of a refunctioned snorkeling mask by 3D-print technology as a tight-fitting full face mask for NIV use. (Photo credited and permission given by ISINNOVA®, Rome, Italy.)

modified strategies when applying NIV on a patient infected with COVID as follows: (a) before initiating or terminating NIV use, the patient's mask must be in place, and personal protective equipment must be worn by caregivers; (b) the mask should be sealed as tightly as possible, and should involve a full face mask, perhaps with a temporary lowering of pressure if the leakage is too high, and an antibacterial filter should be used at the ventilator outlet on the inspiratory circuit and after the mask.[7] Hence, a novel design of tight-fitting, full face mask with virus-proof filters at the exhalation exit may help to solve the contradiction of NIV use in hospitals during the COVID-19 pandemic (Figure 1). Of note, several innovative designs have been proposed and applied clinically.[8,9] An important difference is that these modified NIV interfaces may provide a potentially more closed ventilation system, which may be advantageous in preventing aerosolized transmission of COVID-19 to healthcare workers. Nevertheless, it should be emphasized that there is no single interface suitable for all situations, and a sophisticated selection of an appropriate interface by an experienced care team is a key factor in achieving NIV success while minimizing air leakage, maximizing patient comfort, and synchronizing with the ventilator. If patients need near 24-h NIV support for severe respiratory compromises, alternating masks is recommended to prevent pressure sores, and they should be alternated day and night between two ventilators of the same model, so as not to run a ventilator continuously for days.[10]

Furthermore, periodic prone positioning during mechanical ventilation seems beneficial in improving the oxygenation of patients with COVID-19 complicated by acute respiratory distress syndrome.[6] However, this position might be challenging or even risky if applied to patients with NMD, especially for those frequently associated with kyphoscoliosis. Tracheobronchial compression against the vertebral bodies can occur in patients with severe thoracic kyphoscoliosis on prone positioning.[11] Therefore patients with NMD with anatomical peculiarities may influence the options that pulmonologists choose for prolonged prone ventilation.

Conclusively, it should always be kept in mind that COVID-19 is a rapidly evolving field, and the advisory guideline for respiratory management in patients with NMD is frequently subject to revision. There must be close collaboration between neuromuscular specialists and pulmonologists who should support their hospital to define approved devices and care methods.

Author contribution(s)

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Jong-Hau Hsu: Conceptualization; Resources; Writing-original draft; Writing–review & editing.

Conflict of interest statement

The authors declare that there is no conflict of interest.

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Supplemental material

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