Managing the Patient With Neuromuscular Disease and Respiratory Insufficiency

Though the development of physical limitations associated with neuromuscular disease is often apparent to the clinician, the development of neuromuscular-induced respiratory muscle weakness can be insidious. Respiratory muscle weakness is often overlooked by the clinician, particularly when weakness of the extremities limits physical exertion and no respiratory compensation for exertion is required. Patients with neuromuscular disease are often not found to have respiratory insufficiency until they present in the emergency department with acute respiratory failure secondary to pulmonary congestion resulting from respiratory infection and inadequate cough clearance. Early symptoms of neuromuscular respiratory insufficiency are more often associated with the onset of sleep-disordered breathing.1 On finding symptoms of neuromuscular weakness the clinician should question the patient about symptoms of sleep-disordered breathing and developing dyspnea when supine or with exertion. A comprehensive neuromuscular respiratory evaluation should also be done as a baseline for serial assessment of chronic progressive insufficiency, to support preventive out-patient respiratory care.2,3

In this issue of Respiratory Care, Kelly et al4 describe a rare adult-onset presentation of nemaline myopathy, a muscle disease that can affect the respiratory muscles. In this case the patient was referred by his primary care provider to a respiratory clinic after developing cough and dyspnea on exertion. Although the patient presented with chronic hypercapnic respiratory failure, the immediate use of noninvasive ventilation (NIV) relieved his symptoms and reversed the chronic alveolar hypoventilation, as evidenced by improved arterial blood gas values.

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Though NIV has been more widely used to treat obesity hypoventilation and central and complex sleep-disordered breathing, it could be argued that patients with neuromuscular disease and respiratory insufficiency receive the greatest benefit from NIV. NIV is effective in providing intermittent, often nocturnal, ventilatory support in various neuromuscular diseases,5 and improves survival and quality of life in patients with amyotrophic lateral sclerosis.6

The key to effective NIV for respiratory insufficiency is to provide adequate pressure support to augment respiratory-muscle-induced hypoventilation. “Wide-span” bi-level pressure support (ie, in which the difference between the applied inspiratory and expiratory pressures is ≥ 10 cm H2O) effectively augments ventilation. A minimal expiratory pressure—only that sufficient to flush exhaled gas from the circuit—is better tolerated by patients with neuromuscular disease, and is generally sufficient when neuromuscular weakness does not affect upper-airway patency during sleep. The need for a spontaneous/timed mode with an adequate backup rate is often overlooked when prescribing NIV for nocturnal hypoventilation associated with neuromuscular disease. During rapid-eye-movement (REM) sleep, all of the muscles of ventilation except for the diaphragm develop a state of atonia.7 If diaphragm weakness during REM sleep limits the ability to trigger the ventilator, the patient may not receive adequate ventilatory support during REM sleep, so a backup rate provides a necessary bridge of ventilatory support during REM sleep.

Particular attention must be paid to determining the best patient-ventilator interface with regard to fit, comfort, and effectiveness.8 A self-directed desensitization protocol can help the patient acclimate to NIV before titrating the inspiratory pressure to achieve optimal pressure support.9

Although oronasal masks are not always well tolerated, particularly by patients with neuromuscular disease and therefore unable to remove the mask, oronasal masks may provide the most effective means of limiting air leak, if nasal mask and chin-strap alternatives are not effective, as often occurs in patients with bulbar weakness. A patient with neuromuscular disease who relies on nasal mask as the primary NIV interface should also have an oral-nasal mask alternative in the event of nasal congestion from allergic rhinitis or upper-respiratory infection.

Monitoring and managing mask air leak is particularly important in maximizing the effectiveness of NIV.10 A data-storage device in a bi-level-pressure ventilator can be very helpful for evaluating ongoing mask leak and monitoring the effectiveness of ventilation and patient adher-
ence to NIV. Most neuromuscular disease processes are chronic and progressive and therefore require increasing pressure support to maintain adequate ventilation. An integrated heated humidifier should be prescribed with the bi-level-pressure ventilator, and the patient can adjust the humidification according to comfort with the increasing pressure support.

In their patient, Kelly et al. observed improved forced vital capacity following initiation of NIV. Although not described in their report, a regimen of hyperinflation therapy, either via pressure pre-set or volume insufflation, can reverse atelectasis, improve lung compliance, and help maintain the respiratory health of patients with neuromuscular disease and chronic hypoventilation.11

Correcting neuromuscular-induced hypoventilation is usually the clinician’s primary goal, and providing support to compensate for limited or ineffective cough strength is often overlooked. This is an important component in the overall preventive respiratory care plan for patients with neuromuscular disease. Peak expiratory cough flow (a measure of cough strength) should be evaluated in any comprehensive neuromuscular respiratory assessment.12,13 Cough-augmentation techniques, including mechanical in-exsufflation and manual hyperinflation with assisted cough maneuvers, help patients who have inadequate cough clearance.14,16 In the patient described by Kelly et al. there was no evidence of airway-clearance limitation. This is most likely due to the absence of bulbar involvement and relatively well preserved expiratory muscle strength, as indicated by the patient’s peak expiratory pressure (86 cm H₂O).

The overall management of neuromuscular respiratory insufficiency, including diagnosis and initiation and management of ventilation and cough-augmentation therapies, is within respiratory therapists’ scope of practice. With a strong understanding of neuromuscular respiratory pathophysiology the respiratory therapist is in a unique position to complement the pulmonologist and neurologist in managing the ongoing respiratory care of patients with neuromuscular respiratory insufficiency.

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References