Neuromuscular Disorders and Sleep in Critically Ill Patients

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KEYWORDS

• Neuromuscular disorders • Sleep • Sleep disorders • Critical care
• Intensive care unit • Management

KEY POINTS

- Sleep problems, especially sleep-disordered breathing (SDB), are frequent in neuromuscular patients and contribute significantly to morbidity and mortality.
- SDB usually manifests before any daytime respiratory symptoms evolve in patients with neuromuscular disorders.
- Nocturnal hypoventilation is particularly common, and obstructive sleep apnea and central sleep apnea are also common comorbidities in neuromuscular patients.
- During rapid eye movement (REM) sleep, respiration depends on diaphragmatic effort, and REM-related hypoventilation and SDB are early manifestations in neuromuscular patients with evolving diaphragmatic weakness.

Neuromuscular disorders are frequently associated with sleep-disordered breathing (SDB) abnormalities, although the cumulative prevalence is not well known and probably underestimated. Between 27% and 62% of children[1–3] and 36% and 53% of adults[4] with neuromuscular disorders have SDB, depending on the type of neuromuscular disorder involved, definition of respiratory impairment, and tools used to

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measure SDB. Forty percent of patients followed at a Mexican neuromuscular clinic had sleep or SDB abnormalities.[4] SDB with or without nocturnal hypercapnic hypoventilation is a common complication of respiratory muscle weakness in childhood neuromuscular disorders. SDB was found in 35 of 49 patients (71%), and 24 (49%) had SDB with nocturnal hypercapnic hypoventilation.[3] Patients suffering from neuromuscular disorders may present with impairment at the levels of the upper motor neuron, lower motor neuron, nerve roots, brachial plexus, peripheral nerve, neuromuscular junction, or muscle, causing weakness of respiratory muscles that may result in SDB.

SDB often precedes daytime respiratory symptoms and may be the presenting manifestation in patients with neuromuscular disorders. Hence, untreated SDB may result in acute or chronic respiratory failure, the most common cause of morbidity and mortality in up to 80% of patients with neuromuscular diseases.[5] The risk of respiratory infections is increased by impairment of cough because of respiratory muscle or bulbar weakness, and death is frequently due to respiratory failure. In addition to diaphragmatic and respiratory muscle weakness, several other factors mediate disturbed sleep in patients with neuromuscular disorders, including those summarized in **Box 1**.

During sleep, particularly in rapid eye movement (REM) sleep, upper airway resistance increases, while chemosensitivity and skeletal muscle tone decrease (with the exception of the diaphragm), resulting in hypoventilation and leading to hypoxemic and hypercapnic failure. The most common form of SDB in patients with respiratory muscle weakness is hypoventilation due to reduced tidal volume, particularly during REM sleep, but nocturnal desaturation may occur due to nocturnal hypoventilation, periodic apneas and hypopneas, or ventilation/perfusion mismatching resulting from atelectasis in the supine posture. Secondary lung diseases, such as aspiration from pharyngeal muscles, impair deglutition and decrease cough reflex, predisposing to atelectasis and bronchiectasis, leading to long-term pulmonary fibrosis.

SDB leads to significant deterioration in both subjective and objective sleep quality, with specific common objective alterations in sleep architecture and polysomnographic parameters as outlined in **Box 2**.

SDB is more likely to occur in patients with rib cage and spinal deformities, obesity, and craniofacial abnormalities. Because neuromuscular disorders are most vulnerable to oxygen desaturation during REM sleep, suppression of REM sleep may represent a compensatory mechanism. The risk of respiratory infections including pneumonia is also increased, because of impairment of cough and clearing of secretions, given respiratory and/or bulbar weakness.

Box 1
Factors causing sleep-related difficulties in neuromuscular patients

Factors causing sleep disruption

a. Diaphragmatic and respiratory muscle weakness
b. Upper airway and craniofacial weakness
c. Difficulty with secretion clearance
d. Impairment of cough mechanism
e. Limitation of posture/discomfort due to weakness
f. Diminished ventilatory drive