Sleep-Disordered Breathing in Neurologic Conditions

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KEYWORDS

• Sleep-disordered breathing • Neurologic disorders • Obstructive sleep apnea

KEY POINTS

- Sleep-disordered breathing (SDB) is common in neurologic disorders.
- Obstructive sleep apnea, central sleep apnea, and sleep-related hypoventilation can all be seen in neurologic conditions, with certain sleep-related breathing disorders being more common in specific neurologic conditions.
- The underlying cause of SDB is complex and often multifactorial.
- Identification and treatment of SDB is an important aspect of treating patients with neurologic disease.

INTRODUCTION

Sleep-related breathing disorder or sleep-disordered breathing (SDB) encompasses a range of conditions, including central sleep apnea (CSA), obstructive sleep apnea (OSA), and sleep-related hypoventilation or hypoxemic syndromes.[1] For a list of possible presentations of SDB in neurologic disease, see **Box 1**. SDB is common in neurologic conditions that impact the central and/or peripheral nervous systems. Patients with neurologic conditions are at risk for SDB due to a combination of factors such as muscular weakness, damage to areas of the brain that control respiration, use of sedating medications, and, in some cases, weight gain from limited physical activity (**Box 2**). Recognition and treatment of SDB is an important aspect of treating patients with neurologic disease.

NEUROMUSCULAR DISEASES AND BREATHING DURING SLEEP

Disordered breathing during sleep and neuromuscular diseases are intimately linked. Breakdowns in nerves, muscles, and the connections between them may lead to severe compromises in respiratory function. However, each neuromuscular disorder may have a different impact on breathing, given the variance in the location of the dysfunction. This section discusses some of the common neuromuscular conditions and their resultant SDB.

Amyotrophic Lateral Sclerosis

Amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig disease for the well-known New York Yankee first baseman who suffered from the disorder, is a disease of neuron degeneration in the brain, brainstem, and ventral horn of the spinal cord. Symptoms of this often rapidly progressive disorder include weakness, muscle atrophy and fasciculation, spasticity, dysarthria, dysphagia, and respiratory compromise. Dysfunction of the respiratory system occurs on multiple levels, including pharynx, larynx, intercostal muscles, and diaphragm. Thus, neurons supplying the inspiratory, expiratory, and upper-airway muscles are all affected. A variety of causes have been

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Box 1
Possible presentations of SDB in neurologic conditions

- OSA
- Nocturnal stridor
- CSA, Cheyne-Stokes
- Sleep-related hypoventilation
- Mixed OSA and CSA
- Acquired central alveolar hypoventilation

proposed for this disease, including toxic exposure, genetic disorders, and autoimmune conditions, although no single cause for the disease has been identified.[2] Approximately 15 new cases are diagnosed per day (5600 per year) with an estimate of 30,000 patients currently diagnosed with the disease in the United States at any given time.[3]

Sleep complaints are common in ALS. In one study, 59% of subjects with ALS had sleep complaints compared with 36% of controls. Common sleep complaints include nocturia, sleep fragmentation, and nocturnal cramping.[4] Although not all of the sleep complaints are likely related to underlying SDB, it seems likely that at least some of the symptoms have a respiratory-related underpinning.

In late stages of ALS, patients will have daytime respiratory problems; however, patients will often have nocturnal breathing disturbance much earlier due to the vulnerability of the respiratory system during sleep. In particular, diaphragmatic dysfunction leads to reduced ventilation during sleep, particularly rapid eye movement (REM) sleep, when other accessory muscles do not lend support to breathing. Early intervention with noninvasive ventilation in patients suffering from ALS has improved their quality of life and may also prolong survival.[5,6] Several types of abnormal sleep-related breathing disorders have been discovered in ALS, including OSA, CSA, and hypoventilation, with the frequency of reporting widely variable.

Box 2
Potential contributors to SDB in neurologic conditions

- Muscular weakness, including muscles of respiration
- Damage to respiratory control areas of brainstem
- Sedating medications
- Obesity

Most common of the sleep-related breathing disturbances in ALS is hypoventilation. Because the disease affects the diaphragm, intercostal muscles, and accessory muscles of respiration, the patient moves less air in and out of the lungs (reduced tidal volume). REM sleep is a period of particularly notable hypoventilation. Human REM-related respiration is nearly completely dependent on diaphragmatic movement, which is limited by motor neuron disease. Nocturnal hypoventilation may be difficult to unmask initially, with subtle symptoms that might include restless sleep, insomnia, and morning headaches. As the hypoventilation worsens, it causes frequent arousals from sleep, leading to both nocturnal symptoms of insomnia and daytime symptoms of sleepiness. For a more complete list of symptoms of hypoventilation, see **Box 3**.

Myasthenia Gravis and Lambert Eaton Myasthenic Syndrome

Myasthenia gravis (MG) is characterized by episodic weakness related to the fatigability of voluntary muscles. This autoimmune disorder is defined by prevention of neuromuscular transmission due to blockage of postsynaptic acetylcholine receptors. Repetitive use of the voluntary muscles will cause weakness; rest restores normal function.[7] There are approximately 36,000 to 60,000 cases of MG in the United States, with an estimated prevalence of 14 to 20 cases per 100,000

Box 3
Symptoms of nocturnal hypoventilation

- Air hunger
- Snoring
- Choking
- Orthopnea
- Cyanosis
- Restlessness
- Insomnia
- Daytime hypersomnolence
- Morning headaches
- Drowsiness
- Fatigue
- Depression
- Impaired cognition

Adapted from Perrin C, Unterborn JN, Ambrosio CD, et al. Pulmonary complications of chronic neuromuscular diseases and their management. Muscle Nerve 2004;29(1):5–27; with permission.