Sleep-disordered Breathing in Neuromuscular Disease.

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Abstract
Sleep-disordered breathing in neuromuscular diseases is due to an exaggerated reduction in lung volumes during supine sleep, a compromised physiologic adaptation to sleep, and specific features of the diseases that may promote upper airway collapse or heart failure. The normal decrease in the rib cage contribution to the tidal volume during phasic REM sleep becomes a critical vulnerability, resulting in saw-tooth oxygen desaturation possibly representing the earliest manifestation of respiratory muscle weakness. Hypoventilation can occur in REM sleep and progress into non-REM sleep, with continuous desaturation and hypercarbia. Specific characteristics of neuromuscular disorders, such as pharyngeal neuropathy or weakness, macroglossia, bulbar manifestations, or low lung volumes, predispose patients to the development of obstructive events. Central sleep-disordered breathing can occur with associated cardiomyopathy (e.g., dystrophies) or from instability in the control of breathing due to diaphragm weakness. Mitigating factors such as recruitment of accessory respiratory muscles, reduction in REM sleep, and loss of normal REM atonia in some individuals may partially protect against sleep-disordered breathing. Noninvasive ventilation, a standard-of-care management option for sleep-disordered breathing, can itself trigger specific sleep-disordered breathing events including air leaks, patient-ventilator asynchrony, central sleep apnea, and glottic closure. These events increase arousals, reduce adherence, and impair sleep architecture. Polysomnography plays an important role in addressing pitfalls in the diagnosis of sleep-disordered breathing in neuromuscular diseases, identifying sleep-disordered breathing triggered by noninvasive ventilation, and optimizing noninvasive ventilation settings.

KEYWORDS: hypoventilation; neuromuscular diseases; noninvasive ventilation; polysomnography; sleep-disordered breathing

PMID: 25723731 DOI: 10.1164/rccm.201412-2224CI

[Indexed for MEDLINE]