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Respiratory disorders in neurologic diseases

ABSTRACT

Various neurologic diseases such as multiple sclerosis and Parkinson disease can cause pulmonary complications. Pulmonary disorders often manifest late in a neuromuscular disease, but occasionally a respiratory problem may be the first sign. Often the first signs are sleep disturbances and nocturnal desaturation. Although the diseases are diverse, common principles apply in their management.

KEY POINTS

In neurologic diseases, the forced vital capacity is a simpler and more sensitive marker of restrictive pulmonary impairment than is total lung capacity and is an important tool for follow-up.

Adequate pulmonary management should aim to support inspiratory and expiratory muscle function, as this has been found to decrease hospitalization rates and improve survival.

Noninvasive positive-pressure ventilation via a nasal mask is often effective for the support of inspiratory muscle function, though it may be contraindicated in some patients.

Other treatments include respiratory muscle training and theophylline in select patients.

To avoid worsening hypercapnia, oxygen supplementation should probably be limited to patients already on some form of ventilatory support, or to patients in whom hypoventilation has been excluded.

MANY NEUROLOGIC DISEASES, including multiple sclerosis and Parkinson disease, can progress to the point where they compromise respiratory function (TABLE 1).¹⁻²³ Although the diseases have variable causes and clinical courses, in many cases it is a pulmonary complication that is the cause of death or other serious adverse event.

This review addresses the pathophysiology of pulmonary impairment in diverse neurologic diseases and reviews the clinical evaluation, the role of pulmonary function testing, and management, including medical therapy and support of inspiratory and expiratory function.

PATHOPHYSIOLOGY

Central respiratory control may be normal

Patients with neuromuscular diseases tend to have either a normal or increased central respiratory drive.²⁴⁻²⁷ However, patients who develop nocturnal hypercapnia may have a reduced central drive and reduced daytime ventilatory responses, possibly because progressive accumulation of CO₂ in the cerebrospinal fluid (CSF) raises the pH of the CSF in relation to the partial pressure of CO₂ (P(CO)₂ of the CSF.²⁸

Abnormalities in central respiratory control may play a more direct role in the pathogenesis of respiratory dysfunction in central neurologic diseases such as multiple sclerosis and Parkinson disease.

Howard et al²⁹ found that 6 of 19 patients with multiple sclerosis and respiratory complications had abnormal respiratory control. Patients with multiple sclerosis can develop voluntary or autonomic respiration, diaphrag-

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