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Pulmonary rehabilitation in neuromuscular disorders and spinal cord injury

Abstract

Most patients with impairment of pulmonary function can be differentiated into those who have primarily oxygenation impairment with hypoxia due to predominantly intrinsic lung/airways disease and for whom hypercapnia is an end stage event, and those with lung ventilation impairment on the basis of respiratory muscle weakness for whom hypercapnia causes hypoxia. This distinction is important because, although many patients in the former category have been described to benefit from noninvasive ventilation in the acute care setting, long term use is more controversial. Patients with primarily ventilatory impairment, on the other hand, can benefit from the use of both inspiratory and expiratory muscle aids; and often avoid having any episodes of respiratory failure despite total respiratory muscle paralysis, do not require tracheostomy, and have excellent prognoses with long term home mechanical ventilation.

Ventilatory muscle failure is defined by the inability of the inspiratory and expiratory muscles to sustain one's respiration without resort to ventilator use. Patients with ventilatory muscle failure do not have unlimited breathing tolerance and require ventilatory support and other respiratory muscle aids.

Inspiratory and expiratory muscle aids are devices and techniques that involve the manual or mechanical application of forces to the body or intermittent pressure changes to the airway to assist inspiratory or expiratory muscle function. The most important inspiratory aid is to receive air under pressure when one inhales (intermittent positive pressure ventilation or IPPV). The most important expiratory aid is to have a negative pressure (vacuum) applied to the airway via the nose and mouth when one coughs along with a manual thrust to the belly to further increase cough flows. Illness and death in people with generalized wea-

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kness, such as patients with neuromuscular disease and high spinal cord injury, is almost always due to respiratory difficulty that occurs because of a weak cough. Breathing (inspiratory), expiratory, and throat (bulbar) muscles are needed for effective coughing. The latter are predominantly the abdominal muscles. Clearing airway secretions can be a continual problem but it most often occurs during chest infections.
The following review will describe the most important aspects of pulmonary rehabilitation in patients with muscle weakness /paralysis and its main goals of respiratory muscle substitution to avoid ventilatory failure and promote quality of life in these patients.

Introduction

Ventilatory impairment is either due to central hypoventilation or respiratory muscle dysfunction in neuromuscular disease (NMD). Dysfunction results, most commonly, from weakness, contracture, or myotonia. As for the skeletal muscles, it is both the combination of weakness and contractures that result in dysfunction and this causes respiratory morbidity and mortality when untreated.
A typical example of ventilatory insufficiency is that of the hypercapnic patient with Duchenne muscular dystrophy (DMD) who has normal SpO_2 when awake and who is minimally symptomatic with little or no ventilator use. Patients symptomatic from ventilatory insufficiency most often have dips in SpO_2 below 95%. Symptomatic hypercapnic patients benefit from the use of noninvasive ventilation for at least part of the day, most often overnight. With progressive ventilatory muscle weakness, withdrawal of periods of daily or nightly aid for these patients will eventually result in ventilatory failure.
Neuromuscular disease patients with primarily ventilatory impairment, respiratory morbidity and mortality are a direct result of failing to assist inspiratory and expiratory muscle function as needed. For patients with primarily oxygenation impairment, the respiratory muscles, although not primarily involved, can be placed at a mechanical disadvantage by the development of lung and chest wall deformities, weakened by malnutrition and overuse, and strained to their limits by the need to ventilate stiff, noncompliant, diseased lungs or irreversibly obstructed airways. Overwork, relative or absolute, can eventually lead to secondary respiratory muscle dysfunction and overt respiratory failure. Patients with primarily ventilatory impairment who also have bulbar muscle dysfunction so severe that maximum assisted CPF are less than 160 L/m have essentially irreversible upper airway obstruction.
Ventilatory insufficiency and impaired airway secretion clearance are common