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MANAGEMENT OF PULMONARY COMPLICATIONS IN NEUROMUSCULAR DISEASE

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Normal breathing depends on the function of the ventilatory pump, which consists of the central respiratory control centers, the bony rib cage, diaphragm, and the intercostal and accessory muscles. Normal arterial blood gases and defense of the lung through cough depend on intact muscle function. A wide variety of neuromuscular disorders result in dysfunction of the ventilatory pump that in turn can lead to respiratory failure, pneumonia, and even death. Breathing disorders are recognized as the leading cause of mortality in neuromuscular disease.[9, 25] Appropriate intervention prevents complications and prolongs life in individuals whose neuromuscular disease affects their respiratory system.

ETIOLOGY OF RESPIRATORY FAILURE IN NEUROMUSCULAR DISEASE

Respiratory failure in neuromuscular diseases results from a number of factors, including: (1) respiratory muscle weakness and fatigue; (2) alteration in respiratory system mechanics; and (3) impairment of the central control of respiration.

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Respiratory Muscle Weakness and Fatigue

Respiratory muscle weakness and fatigue are frequent contributors to ventilatory failure in the patient with neuromuscular disease. Respiratory muscle *weakness*, the inability of the respiratory muscles to generate normal levels of pressure and flow during inspiration or expiration, occurs because of lack of appropriate stimulation of muscle fibers, as in spinal cord injury, or intrinsic dysfunction of the fibers themselves, as in the muscular dystrophies. Respiratory muscle *fatigue*, defined as the failure of a muscle to maintain a constant force during repetitive contractions[55] can occur because of weakness of the respiratory muscles themselves or because of an increase in load. Progressive muscle weakness and fatigue lead to restrictive lung disease and ultimately to hypoventilation, hypercarbia, and respiratory failure. Respiratory muscle fatigue and ventilatory failure usually occur only with significant impairment of respiratory muscle strength, at a level less than 30% of predicted values.[10]

Alteration in Respiratory Mechanics

In addition to effects on muscle contraction, neuromuscular disease increases elastic and resistive loads on the respiratory muscles. Both types of loads increase the work of breathing and hasten ventilatory failure. Increases in elastic loads are a consequence of an increase in stiffness of both the lungs[9] and the chest wall.[21, 42] Airway secretions and an ineffective cough mechanism for their clearance result in increased airways resistance. Finally, kyphoscoliosis, frequently associated neuromuscular disease processes, can increase the work of breathing. Deformation of the thoracic cage further increases chest wall stiffness and produces mechanical misalignment of the respiratory muscles, thus lessening their ability to operate effectively against the increased elastic and resistive loads.[9]

Impairment of Control of Ventilation

Disorders of central control of respiration frequently are associated with neuromuscular disease processes. Defects in control of respiration may be caused by the disease itself, as in bulbar poliomyelitis, or secondary to hypoxemia and hypercarbia associated with a disease process as it progresses, as in Duchenne muscular dystrophy. The first indicators of disordered respiratory control may occur in association with sleep. Significant nocturnal decreases in partial pressure of oxygen (Pa_{O_2}) as well as elevations in arterial partial pressure of carbon dioxide (Pa_{CO_2}) have been reported.[29, 30, 43] These changes are most likely to occur during rapid eye movement sleep when a pattern of rapid shallow breathing develops.[30, 43] This hypoventilation sometimes is associated with apneic episodes, leading to further hypercapnia.[3, 29, 30] Hypercapnia or hypoxemia occurring at night may have a role in reducing daytime central respiratory drive by depressing central drive centers directly and also by increasing the "bicarbonate pool."[29, 30, 45] This latter effect blunts the stimulus to breathe generated by respiratory acidosis and perpetuates the hypercapnic state.

Presentation of Respiratory Failure

Respiratory failure will present in one of three ways in patients with neuromuscular disease: (1) as acute respiratory failure such as that occurring in high level spinal cord injury; (2) as an acute respiratory decompensation in a chronic disease such as amyotrophic lateral sclerosis (ALS); or (3) as a chronic worsening of a gradually progressive disease such as Duchenne muscular dystrophy (DMD).

Treatment of acute respiratory failure frequently involves endotracheal intubation and positive pressure ventilation in the ICU. Many times tracheostomy tube placement is required. In this situation, unfortunately, there are few choices of methods of ventilation. It is possible, however, to wean a patient from invasive positive pressure ventilation by utilizing a variety of noninvasive ventilatory techniques.[6] Thus the tracheostomy tube, which some patients find objectionable, is not necessarily permanent.

The insidious onset of respiratory failure in neuromuscular diseases is a common occurrence and provides an opportunity for early intervention with a variety of ventilator options. Noninvasive forms of both positive and negative ventilation as well as the rocking bed and pneumobelt have been used effectively in reversing, at least temporarily, progressive chronic respiratory failure.[19, 24, 32, 36] Initially, patients may require ventilatory support for only part of the day. In these cases, nocturnal ventilatory support has been shown to be of great value.[15, 19, 20, 32, 36, 53] Daytime ventilation either full-time or for prescribed periods can be utilized as muscular weakness progresses.

METHODS OF VENTILATORY SUPPORT IN NEUROMUSCULAR DISEASE

Mechanical Ventilation

Artificial ventilation as a means of prolonging human life has been referred to since Biblical times. More recently, but as early as the mid-nineteenth century, ventilation during surgery, including open-chest operations, has been possible. It was not until 1927 and the invention of the motorized iron lung by Drinker[18] that long-term mechanical ventilation became a possibility. Chronic ventilation via a tracheostomy tube became possible in the 1950s with the development of reliable, smaller positive pressure ventilators. Today a wide variety of devices exist for chronic ventilation of patients with neuromuscular disease as well as other forms of respiratory failure.

DEVICES AVAILABLE FOR MECHANICAL VENTILATION OF NEUROMUSCULAR PATIENTS

I. NEGATIVE PRESSURE VENTILATORS
   a. Full body ventilator (tank ventilator or iron lung)
   b. Raincoat ventilator ("poncho" or "pneumowrap")
   c. Cuirass ventilator (chest shell)
   d. Pneumosuit ventilator with leggings
II. POSITIVE PRESSURE VENTILATORS
   a. Via tracheostomy
   b. Noninvasive
      1. Via full face mask
      2. Via nasal mask
      3. Mouthpiece with lipseal
III. VENTILATORS RESULTING IN PASSIVE MOVEMENT OF THE DIAPHRAGM
   a. Pneumobelt
   b. Rocking bed
IV. PHRENIC NERVE PACING

Negative Pressure Ventilators

Negative pressure ventilators have been used for many years, bringing extensive experience ventilating patients with a wide range of neuromuscular diseases. Negative pressure ventilators function by applying a negative pressure to the surface of the thorax and abdomen, expanding the chest wall and lungs and thus promoting the movement of air into the lungs. Exhalation occurs passively because of the inward elastic recoil of the chest wall and lungs. Because pleural pressure is lowered during inspiration, this type of ventilation more closely mimics spontaneous breathing than does positive pressure ventilation. Full body ventilators such as the iron lung encase the patient's entire body except for the head, which remains outside the device and is sealed at the neck with a rubber or plastic collar. Cyclic negative pressure is created within the tank by a bellows pump connected to an electric motor that can be powered by alternating current or a back-up battery supply. Respiratory rate as well as the volume of each breath can be adjusted. Because of the large size and weight (325 kg)[33] of the iron lung, smaller and lighter devices have been developed. The Portalung is a fiberglass shell that weighs only 1/6 as much as the iron lung and is powered by a separate, large volume negative pressure generator. One drawback to both of these devices, particularly the iron lung, is lack of access to the patient.

In addition, some patients become claustrophobic when these devices are used.

The cuirass or chest shell (Fig. 1) is a device that was developed to overcome the portability and confinement problems of the larger tank ventilators. It consists of a fiberglass shell that fits over the anterior chest and abdomen. This is attached to a negative pressure generator via 2″ pressure tubing. Intermittent negative pressure is generated within the shell, resulting in expansion of the chest wall and lungs. Although portable and more convenient than the "full-body" negative pressure ventilators, the cuirass is the least efficient of the negative pressure ventilator devices.[13] This inefficiency in part may be a consequence of improper fit of the device to the patient's thorax and abdomen, resulting in significant air leaks, which often occur with mass-produced cuirasses. This problem may be circumvented if the cuirass is custom designed and manufactured individually, unfortunately at considerable expense.

The light weight and ease of application are attractive features of this device. Although the cuirass has been used for patients who require 24-hour ventilation, it probably is best reserved for those who require less ventilator assistance.

The pneumowrap negative pressure ventilator[52] is a poncho-like rubberized garment worn over a shell-like grid placed on the patient's chest. Drawstrings or Velcro attached to the wrist, head, and lower waist openings seal the pneumowrap over the patient. A negative pressure generator attached via 2″ tubing to the poncho allows generation of negative pressure around the chest and abdomen. The grid is essential to provide a gas volume around the chest that can be decompressed, causing the thorax to expand. This device does not have to be customized and is relatively inexpensive. Unfortunately, in many patients significant air leaks around the legs can result in inefficient ventilation.[35]

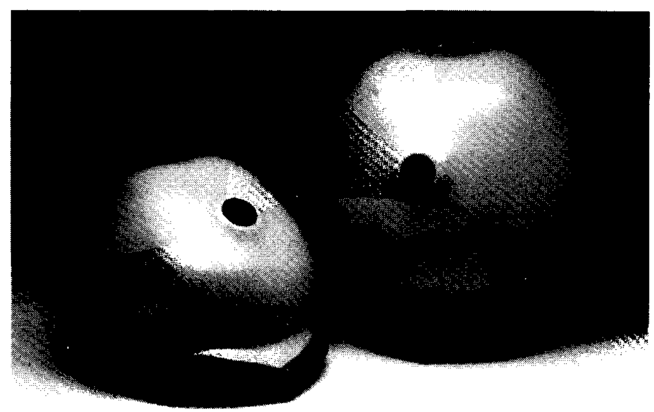

Figure 1. Cuirass or "chest-shell" device for application of negative pressure ventilation. The device (two sizes shown) fits over the anterior thorax and abdomen and is held in place with Velcro straps.