Noninvasive Positive Pressure Ventilation in the Treatment of Hypoventilation in Children

Iris A. Perez, MD[a,b], Thomas G. Keens, MD[a,b], Sally L. Davidson Ward, MD[b,c,*]

KEYWORDS

- Hypoventilation • Chronic respiratory failure
- Noninvasive ventilation • NPPV
- Bilevel positive airway pressure • Neuromuscular disease
- Ventilatory muscle weakness

Adequate ventilation requires sufficiently robust central respiratory control and ventilatory muscle function to overcome the workload imposed by the pulmonary mechanics and properties of the upper airway. Hypoventilation due to chronic respiratory failure (CRF) in childhood therefore can result from dysfunction of each of these components of the respiratory system. Noninvasive positive pressure ventilation (NPPV) can provide ventilatory support for children with hypoventilation or CRF caused by a variety of underlying clinical entities. Children with CRF stemming from central respiratory control abnormalities, ventilatory muscle weakness or, in some circumstances, restrictive or obstructive lung disease can be managed successfully with NPPV. Although NPPV offers the advantage of not requiring a tracheostomy with the attendant complications, it is not appropriate for all children with CRF because use is generally not well tolerated during wakefulness and the devices have limited portability. Successful use requires cooperation on the part of the child, thus making introduction of the therapy and selection of the interface critical for adherence. Management of CRF requires a multidisciplinary approach to educate and support the patient and family, and to ensure patient safety and quality of life. When NPPV is used for CRF caused by a progressive disease, periodic retitration of pressures is required as well as a consideration of the ongoing appropriateness of noninvasive ventilation. Although NPPV has wide applicability in the pediatric intensive care unit setting for acute respiratory failure, this topic is beyond the scope of this discussion.

PHYSIOLOGY OF CHRONIC RESPIRATORY FAILURE IN CHILDREN

The ability to sustain spontaneous ventilation requires adequate function of neurologic control of breathing, ventilatory muscles, and lung mechanics. Significant dysfunction of any of these 3 components of the respiratory system may impair a child's ability to breathe spontaneously. Respiratory failure occurs when central respiratory drive and/or ventilatory muscle power are inadequate to overcome the respiratory load (**Fig. 1**).

[a] Childrens Hospital Los Angeles, 4650 Sunset Boulevard, Los Angeles, CA 90027, USA
[b] Keck School of Medicine, University of Southern California, Los Angeles, CA, USA
[c] Division of Pediatric Pulmonology, Childrens Hospital Los Angeles, Mailstop #83, 4650 Sunset Boulevard, Los Angeles, CA 90027, USA
* Corresponding author. Division of Pediatric Pulmonology, Childrens Hospital Los Angeles, Mailstop #83, 4650 Sunset Boulevard, Los Angeles, CA 90027.
E-mail address: sward@chla.usc.edu

Sleep Med Clin 5 (2010) 471–484
doi:10.1016/j.jsmc.2010.05.013
1556-407X/10/$ – see front matter

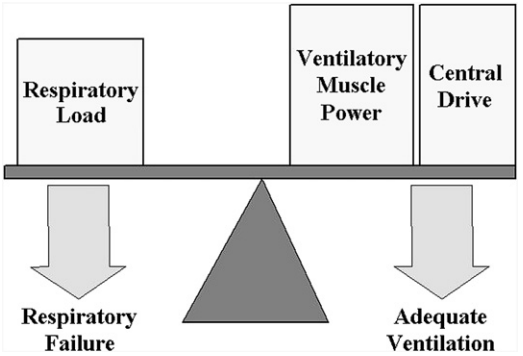

Fig. 1. The respiratory balance. In normal individuals, ventilatory muscle power and central drive are more than adequate to overcome the respiratory load, tipping the balance to the right, which results in adequate ventilation. However, when ventilatory muscle power and/or central drive are sufficiently decreased and/or the respiratory load is sufficiently increased, or some combination thereof, ventilatory muscle power and central drive may not be sufficient to overcome the respiratory load. The balance will tip to the left, and respiratory failure will result.

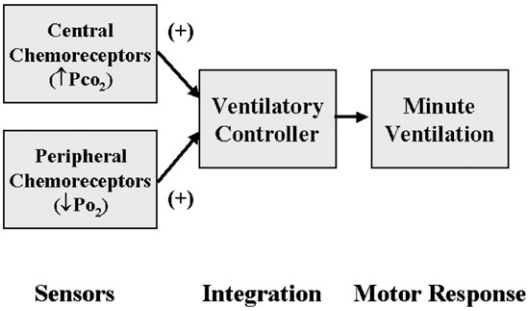

Fig. 2. Neurologic control of breathing. Neurologic control of breathing can be divided into sensors, integration, and motor response. The most important sensors are central and peripheral chemoreceptors, which sense CO_2 and oxygen, respectively. This information is integrated in the brainstem and other areas of the brain. The motor response is minute ventilation or breathing.

CRF occurs if the cause of this imbalance is not reversible, and chronic ventilatory support will therefore be required.[1–6]

CRF implies that a chronic, perhaps irreversible, underlying respiratory disorder is causing respiratory insufficiency that results in inadequate ventilation or hypoxia.[1] The diagnosis of CRF is usually made once repeated attempts to wean from assisted ventilation have failed for at least 1 month in a patient without superimposed acute respiratory disease, or in a patient who has a diagnosis with no prospect of being weaned from the ventilator. This article focuses on CRF, which means that the patient has been determined to be ventilator dependent, and that he or she cannot be weaned from assisted ventilation at the near future.

CENTRAL HYPOVENTILATION SYNDROMES

The cause of CRF in children with central hypoventilation syndromes is inadequate central respiratory drive, either congenital or acquired.[7–13] The congenital form may be genetic (congenital central hypoventilation syndrome) or result from an identifiable brainstem lesion.[7,11–16] Acquired forms of central hypoventilation syndrome may be caused by brainstem trauma, tumor, hemorrhage, stroke, or infection.[8,16]

The primary components of respiratory control are oxygen and CO_2 sensors, integration of input from receptors, and a motor response (**Fig. 2**). The central chemoreceptor, located in the medulla, is sensitive to changes in P_aCO_2. CO_2 in arterial blood diffuses across the blood-brain barrier, increasing H^+ concentration in cerebral spinal fluid (CSF). Central chemoreceptor cells respond to changes in CSF H^+. This mechanism comprises a dynamic system sensitive to small PCO_2 changes. Minute ventilation increases linearly with increasing P_aCO_2. Therefore, central chemoreceptors are responsible for breathing minute to minute. Blood H^+ and HCO_3^- ions do not readily diffuse across the blood-brain barrier. However, with time, if PCO_2 remains chronically elevated, resulting in a chronic elevation of CSF H^+, renal conservation of HCO_3^- will determine a new CSF H^+ baseline. In this way, central chemoreceptors may habituate to high PCO_2 values, as evidenced by a normal arterial pH in the presence of an elevated P_aCO_2. Peripheral chemoreceptors, located in the carotid bodies at the bifurcation carotid arteries, are sensitive to changes in P_aO_2, large changes in pH, and large changes in P_aCO_2. Peripheral chemoreceptor function is tied closest to the hypoxic ventilatory response. Minute ventilation increases exponentially as P_aO_2 decreases and linearly as S_aO_2 decreases. Central and peripheral chemoreceptors act synergistically so that in a patient with both hypercapnia and hypoxia, ventilation will be stimulated more than by hypercapnia or hypoxia alone. Note that central and peripheral chemoreceptors are in anatomically distinct sites. Therefore, damage to the brainstem does not affect afferent peripheral chemoreceptor function.

Based on the function of the chemoreceptors, one can predict the clinical picture associated with central hypoventilation syndromes due to isolated chemoreceptor dysfunction. Patients with