Noninvasive Positive Pressure Ventilatory Support Begins During Sleep

John R. Bach, MD[a,b,*]

KEYWORDS

• Noninvasive ventilation • Noninvasive ventilatory support
• Continuous noninvasive intermittent positive pressure ventilatory support
• Continuous positive airway pressure • Mechanical insufflation-exsufflation • Ventilatory pump failure
• Neuromuscular disease

KEY POINTS

- Patients with weak or paralyzed respiratory muscles should only be prescribed and use sleep nasal NVS when symptomatic without it and be switched from nasal to mouthpiece interfaces for daytime NVS as needed.
- Volume rather than pressure preset ventilation is used for patients who can air stack.
- Bilevel PAP used at high (ventilatory support) spans is reserved only for patients who cannot afford portable ventilators.
- Because many CNVS-dependent patients with SMA type 1 have no bulbar-innervated muscle function, clearly tracheostomy tubes are unnecessary for respiratory support even in the total absence of bulbar-innervated muscle function unless central nervous system/upper motor neuron hypertonicity results in inadequate upper airway patency for MIE to clear airway secretions as necessary.
- Of all the neuromuscular diseases, insufficient upper airway patency for effective mechanical in-exsufflation and, therefore, eventual need for tracheotomy only occurs for patients with ALS.
- At least for patients with DMD for which a large historically controlled study is available, patients live 10 years longer using CNVS than TMV.

Sir Patrick: Don't misunderstand me, my boy, I'm not belittling your discovery. Most discoveries are made regularly every 15 years; and it's fully a hundred and fifty since yours was made last. That's something to be proud of…

—George Bernard Shaw in "The Doctor's Dilemma" 1906, a novel about the moral dilemmas created by limited resources and the conflicts between the demands of private medicine as a business and as a vocation. Things have not changed very much.

The author has no commercial or financial conflicts of interest to report.
This work was not supported by any external funding.
[a] Department of Physical Medicine and Rehabilitation, Rutgers University New Jersey Medical School, Behavioral Health Building, F5759, 60 Orange Avenue, Newark, NJ 07103, USA; [b] Center for Mechanical Ventilation Alternatives and Pulmonary Rehabilitation, University Hospital, 150 Bergen Street, Newark, NJ 07103, USA
* Center for Mechanical Ventilation Alternatives and Pulmonary Rehabilitation, University Hospital, 150 Bergen Street, Newark, NJ 07103.
E-mail address: bachjr@njms.rutgers.edu

Sleep Med Clin ■ (2017) ■–■
http://dx.doi.org/10.1016/j.jsmc.2017.07.010
1556-407X/17/

HISTORICAL PERSPECTIVE

Polysomnography and Sleep-Disordered Breathing

Although it was known for 50 years that the brain emitted electrical signals, it was not until 1929 that the German psychiatrist Hans Berger demonstrated electroencephalographic differences between sleep and wakefulness for humans.[1] In 1956 Bickelmann and colleagues[2] defined the pickwickian syndrome. Then in the early 1960s polysomnography including CO_2 measurements was performed for patients with the classic symptoms of sleep-disordered breathing (SDB).[3] In 1965 Gastaut and colleagues[4] reported that obstructive and mixed obstructive/central events rather than hypercapnia were the causes of hypersomnolence and other symptoms. As a result, CO_2 measurements were discontinued in favor of simple nasal airflow. Publications in 1969 and 1970 reported relief of symptoms by tracheostomy, further supporting the idea that airway obstruction was the problem.[5,6] In 1972 Christian Guilleminault introduced the monitoring of cardiorespiratory parameters[7]; and in 1974 Jerome Holland coined the term "polysomnogram."[8] Therefore, despite Charles Dickens' classic description of SDB in his novel David Copperfield in 1850, SDB was essentially "discovered" more than 100 years later.

In 1876 von Hauke[9] of Austria applied continuous negative airway pressure and continuous positive airway pressure (CPAP) using a chest shell. He also administered CPAP via an oronasal interface to treat atelectasis.[10] His work was forgotten until the late 1930s when Poulton and Barach independently used CPAP for treating acute pulmonary edema. Then in 1974 CPAP was delivered to 20 newborns with respiratory distress syndrome.[11] In 1981 Sullivan and coworkers[12] published CPAP outcomes for five patients with SDB. Once CPAP interfaces (masks) became commercially available in 1984, along with polysomnography to diagnosis and guide in its treatment, the CPAP treatment paradigm for SDB became widely accepted.

Besides increasing functional residual capacity, CPAP acts as a pneumatic splint to keep the upper airway open so that patients can use their inspiratory muscles to ventilate their lungs. It does not, however, provide ventilatory assistance, that is, support for inadequate inspiratory muscle force such as occurs for those with obesity-hypoventilation syndrome, postpoliomyelitis survivors, spinal cord injury (SCI), critical care neuromyopathy, or neuromuscular disease (NMD). Indeed, the failure of CPAP at any pressure to aid obesity-hypoventilation patients who have both airflow obstruction and insufficient inspiratory muscle force for their increased work of breathing induced Respironics Inc (Murrysville, PA) to develop bilevel positive airway pressure (PAP) and place their "BiPAP-ST machine" on the market in 1990. By permitting the independent adjustment of inspiratory positive airway pressure (IPAP) and expiratory positive airway pressure (EPAP), their machines provide ventilatory assistance as a function of the IPAP EPAP span. Thus, they are used as pressure preset ventilators and can provide full ventilatory support if used at spans of at least 18 cm H_2O or more, but because they had no security alarms or internal or external battery function they have not been sanctioned for use for ventilatory support.

Noninvasive Ventilation Versus Noninvasive Ventilatory Support

The goal of sleep doctors has been to titrate away apneas and hypopneas using noninvasive ventilation (NIV), a term that has become synonymous with CPAP and bilevel PAP at the lowest effective bilevel PAP spans. This is typically the approach used for patients with NMD. As a result the polysomnograms performed on them do not typically include CO_2 monitoring and their symptoms are attributed to central and obstructive events because polysomnographies are programmed to interpret every apnea and hypopnea as being caused by central or obstructive events rather than inspiratory muscle dysfunction. Irrespective of how weak they are, patients with NMD are often prescribed or extubated to bilevel PAP scans less than 10 cm H_2O or inadequate for full ventilatory support.

Because other than for my papers nowhere else in the medical literature do the benefits of NIV include noninvasive ventilatory support (NVS), it is now time to coin a new term and abbreviation. Indeed, I have had a patient with NMD referred to me unsuccessfully attempting to use 23 cm H_2O IPAP and 19 cm H_2O EPAP. It would have been less uncomfortable to breathe in a hurricane. After switching him and other similarly managed patients to volume preset or in his case pressure preset ventilation at 23 cm H_2O with no EPAP or positive end-expiratory pressure (PEEP), their daytime and sleep CO_2 normalized, symptoms were alleviated, and with progressive disease they eventually became dependent on continuous NVS (CNVS) without ever developing acute respiratory failure (ARF), being hospitalized, or needing a tracheostomy tube.