Maintaining an “Oral Tradition”: Specific Equipment Requirements for Mouthpiece Ventilation Instead of Tracheostomy for Neuromuscular Disease

Boitano and Benditt\(^1\) have applied the scientific method to provide further understanding of an aspect of airflow management that is practiced by very few centers—one that is in great need of critical investigation and dissemination. It has been demonstrated that the improvement in nocturnal ventilation provided by noninvasive means carries over into the daytime hours and improves diurnal gas exchange.\(^2\) This improvement, however, may be temporary in patients with progressive neuromuscular diseases such as amyotrophic lateral sclerosis and muscular dystrophy.

Approaches to 24-hour ventilatory needs differ from center to center. Several research groups have reported on patients with neuromuscular disease who use mouthpiece ventilation, even up to 24 hours a day, to assist or replace the function of respiratory muscles.\(^3\)–\(^5\) Further, the American Thoracic Society consensus statement on Duchenne muscular dystrophy identifies mouthpiece ventilation as a successful means of daytime noninvasive breathing support.\(^6\) In our own practice we have identified a number of patients with amyotrophic lateral sclerosis, Duchenne muscular dystrophy, postpolio, or high spinal-cord injury, among other conditions, who have little or no measurable respiratory muscle function but can maintain 24-hour ventilation using mask ventilation at night and mouthpiece ventilation during the day.

Despite this evidence and experience, many patients are routinely subjected to unnecessary tracheostomy when long-term nocturnal noninvasive ventilation proves inadequate for daytime gas exchange. A patient who can adequately protect the airway and has sufficient oral function to hold and seal a mouthpiece can generally manage his or her daytime ventilatory needs. Many clinicians are unaware that this can be achieved by using a portable volume ventilator to provide assistance to weakened or paralyzed respiratory muscles, even if there is no spontaneous respiratory muscle function. Even in a recent comprehensive review of noninvasive ventilation in neuromuscular disease, only passing reference was made to mouthpiece ventilation.\(^7\) Physicians may be familiar only with intensive-care ventilators, whose dual-limb circuits cannot provide open mouthpiece ventilation, because exhaled volume is required to measure minute ventilation and prevent alarming.

Most clinicians prescribe pressure-limited ventilators for sleep, as these allow adjustments to inspiratory time, rise time, leak compensation, and other features that make them ideal for comfortable, nocturnal support. However, these devices are not capable of achieving the higher pressure necessary for effective “breath-stacking” or lung-volume recruitment. Portability for chair-mounting has also been difficult, because of battery limitations. Volume-limited ventilators can provide both the tidal volume for sustained ventilation and the pressure and volume needed for effective lung-volume recruitment and achievement of a maximum insufflation capacity.\(^8\) They are also equipped with batteries and the capability of prolonged mobility.

Although such ventilators have been successful in providing this support and have been reported by several authors, to our knowledge, no systematic evaluation or comparison of available devices has been performed. Boitano and Benditt\(^1\) evaluated 8 home volume ventilators. They determined the specific settings for each ventilator to allow mouthpiece ventilation, and they found that 6 of the 8 ventilators tested were capable of providing mouthpiece ventilation without alarming.

Boitano and Benditt’s Table 2 presented the required flow rates and inspiratory times for all but two of the ventilators.\(^1\) This provides a useful guide to required settings to prevent low-pressure alarming, but these results obviously apply only to the specific circuit and mouthpiece described, and within the tidal volumes indicated. A minimal increase in circuit resistance allows the Puritan Bennett LP10 (Mallinckrodt, Minneapolis, Minnesota) to provide effective mouthpiece ventilation without annoying alarms. In our out-patient clinic we provide mouthpiece ventilation primarily with the Puritan Bennett LP10 and the LTV1000 (Pulmonetic Systems, Minneapolis, Minnesota). In our experience, the Puritan Bennett LP10 will support mouthpiece ventilation with a 15mm angled mouthpiece (Part 1004524, Respironics, Murrysville, Pennsylvania), a tapered flex tube (model 4-000614-00, Mallinckrodt, Minneapolis Minnesota), and disposable circuit (model 6461/6463, Mallinckrodt, Minneapolis Minnesota).

In clinical practice the ventilator breath rate, inspiratory time, and tidal volume are adjusted according to patient comfort. A tidal volume greater than 1000 mL may be necessary to satisfy the patient’s breathing requirement, especially when the ventilator circuit needs further modifications such as flow restrictors, an innovative mouthpiece, and/or when minimal mouth leaks are present.

Boitano and Benditt also described lung-volume recruitment or breath-stacking, retaining serial breaths from the ventilator with a closed glottis. Contrary to their description of lung “hyperinflation,” which implies the introduction of a pathophysiologic or even risky state, the provision of mouthpiece ventilation to enable lung-volume recruitment is a measure to restore the restricted vital capacity toward a more normal volume, not a hyperinflated one. For example, a patient with spinal-cord injury and a vital capacity of 1.0 L (predicted 3.5 L) may be able to perform mouthpiece ventilation and lung-volume recruitment to 3.0 L. This represents a 200% increase in vital capacity, also referred to as the maximum insufflation capacity, but the lungs are not hyperinflated, since they are still below the predicted values. This enables more effective cough and may have an important effect on lung and chest-wall mechanics.

We applaud Boitano and Benditt for endeavoring to bring a clearer understanding of the requirements for mouthpiece ventilation using common home, volume ventilators, and for increasing awareness of this critical support. Additional investigations are required to further elucidate the role of mouthpiece ventilation in the ventilatory management of patients with neuromuscu-
lar disease, particularly in preventing tracheostomy and providing optimum respiratory mechanics and airway clearance.

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REFERENCES