Evaluation of Ventilators for Mouthpiece Ventilation in Neuromuscular Disease

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BACKGROUND: Daytime mouthpiece ventilation is a useful adjunct to nocturnal noninvasive ventilation (NIV) in patients with neuromuscular disease. The aims of the study were to analyze the practice of mouthpiece ventilation and to evaluate the performance of ventilators for mouthpiece ventilation. METHODS: Practice of mouthpiece ventilation was assessed by a questionnaire, and the performance of 6 home ventilators with mouthpiece ventilation was assessed in a bench test using 24 different conditions per ventilator: 3 mouthpieces, a child and an adult patient profile, and 4 ventilatory modes. RESULTS: Questionnaires were obtained from 30 subjects (mean age 33 ± 11 y) using NIV for 12 ± 7 y. Fifteen subjects used NIV for > 20 h/day, and 11 were totally ventilator-dependent. The subject-reported benefits of mouthpiece ventilation were a reduction in dyspnea (73%) and fatigue (93%) and an improvement in speech (43%) and eating (27%). The bench study showed that none of the ventilators, even those with mouthpiece ventilation software, were able to deliver mouthpiece ventilation without alarms and/or autotriggering in each condition. Alarms and/or ineffective triggering or autotriggering were observed in 135 of the 198 conditions. The occurrence of alarms was more common with a large mouthpiece without a filter compared to a small mouthpiece with a filter ($P < .001$), but it was not related to the patient profile, the ventilatory mode, or the type of ventilator. CONCLUSIONS: Subjects are satisfied with mouthpiece ventilation. Alarms are common with home ventilators, although less common in those with mouthpiece ventilation software. Improvements in home ventilators are needed to facilitate the expansion of mouthpiece ventilation. *Key words: daytime mouthpiece ventilation; neuromuscular disease; home ventilators; bench; questionnaire.* [Respir Care 2014;59(9):1–•. © 2014 Daedalus Enterprises]

Introduction

Numerous neuromuscular diseases are characterized by an inevitable decline in respiratory muscle performance, resulting in progressive nighttime and ultimately daytime respiratory failure. Noninvasive ventilation (NIV) is a rec-

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ognized and validated first-line treatment for respiratory failure in these subjects.[1,2] Indeed, NIV normalizes nocturnal gas exchange and initially also daytime gas exchange.[3] However, as respiratory muscle weakness progresses, daytime ventilatory failure may occur, which requires the extension of NIV during the day.

Daytime ventilation can be performed with the interface used at night or a mouthpiece as sip ventilation.[4] Mouthpiece ventilation has been reported to reduce the risk of respiratory infection due to tracheostomy and to improve cough, speech, dyspnea, survival, and patient quality of life.[5-8] Mouthpiece ventilation is not widely used due to a lack of knowledge or information but also to inadequate equipment. Few supports are commercially available. Different mouthpieces can be used, with the choice being determined by the ability of the patient to seal his lips around the mouthpiece during a sip but also by the resistance of the mouthpiece.[9] Indeed, most of the current home ventilators have not been specifically designed for mouthpiece ventilation. Home ventilators have low-pressure alarms that sound when circuit pressure drops, indicating tube disconnection. Thus, open-circuit ventilation cannot be performed because of low-pressure alarms. However, open-circuit mouthpiece ventilation can be realized when sufficient peak inspiratory flow is used to create enough back pressure (2–3 cm H_2O) due to mouthpiece resistance.[9] When the ventilator backup rate in the assist/control mode is sufficient to prevent apnea alarms, the ventilator circuit may normally remain open for extended periods without low-pressure or apnea alarms. The patient may be able to receive a ventilator-assisted breath as often as needed by making a sip effort through the mouthpiece. Only one bench study analyzed the ability of home ventilators to provide mouthpiece ventilation.[9] This study showed that 6 of 8 home ventilators supported mouthpiece ventilation, but all of the ventilators that supported mouthpiece venti-

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QUICK LOOK

Current knowledge

Daytime mouthpiece ventilation is a useful adjunct to nocturnal noninvasive ventilation in subjects with neuromuscular disease. Mouthpiece ventilation improves patient tolerance and reduces fatigue.

What this paper contributes to our knowledge

Patients prescribed daytime mouthpiece ventilation are satisfied with this therapy. Home mechanical ventilators designed for invasive ventilation have frequent alarms during mouthpiece ventilation. Home ventilators with software to promote mouthpiece ventilation have fewer alarms and improve patient satisfaction.

lation maintained a functional low-pressure alarm in the event of a circuit disconnect when set at the respective manufacturer's minimum alarm pressure.

The first aim of our study was thus to analyze the practice of mouthpiece ventilation. The second aim was to evaluate the performance of 6 home ventilators for mouthpiece ventilation.

Methods

Clinical Survey

The survey was part of a large French and Belgium survey on long-term mechanical ventilation in neuromuscular subjects. The general objectives of the survey were (1) to evaluate the subjects' knowledge about and comfort with their long-term mechanical ventilation, (2) to compare subjects' and prescribers' opinions and expectations regarding long-term mechanical ventilation, and (3) to compare the equipment used by the subjects with the prescribers' current mechanical ventilation prescriptions. The detailed description and results of the survey have been published recently.[10]

In brief, data were collected via questionnaires specifically developed for the study. The patient questionnaires were completed by health care professionals between September 2010 and July 2011. A health care professional explained all of the definitions such as synchronization, effectiveness, comfort of ventilation, and others in order to improve the understanding of the questions. The objective was to include at least 200 subjects with an equal distribution between subjects treated with NIV during the night only, subjects treated with NIV during both the night and day, and subjects treated with invasive mechanical ventilation via a tracheostomy. Subjects were recruited in ref-
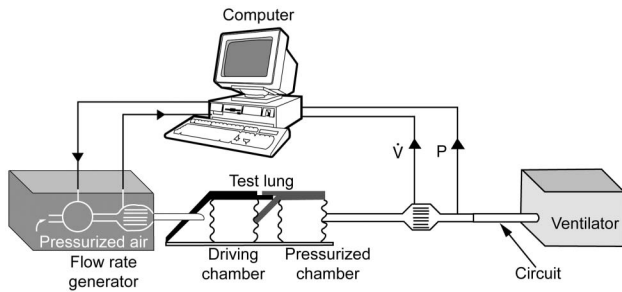
Fig. 1. Bench model used for the study. $\dot{V}$ = air flow; P = airway pressure.
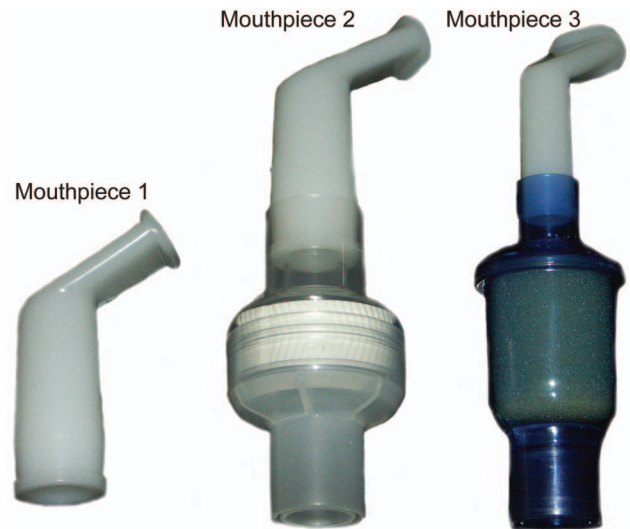

Fig. 2. Mouthpieces and filters used for the bench study. #1: Large rigid plastic mouthpiece (22-mm angled, Philips Respironics). #2: Large rigid plastic mouthpiece (22-mm angled, Philips Respironics) with a filter (Dar heat-and-moisture exchanger, Covidien). #3: Small rigid plastic mouthpiece (15-mm angled, Philips Respironics) with a filter (Hygroflux 1, Vygon, Ecouen, France).

erence medical centers for neuromuscular diseases in Belgium (Inkendaal) and France (Lille, Grenoble, Marseille, Nice, Paris, and Garches) and in residential facilities for neuromuscular subjects in France (Évry, Montpellier, and Angers). The criteria for patient inclusion were (1) diagnosis of a neuromuscular disorder, (2) mechanical ventilation for at least 1 year, and (3) the willingness and availability to participate in the study (an interview of 90 min). The scientific committee decided not to include subjects with amyotrophic lateral sclerosis due to the rapid progression of this disease. For the study, only subjects using mouthpiece ventilation were analyzed.

The study was approved by the Commission Nationale de l'Informatique et des Libertés in agreement with French regulation.

Bench Study

The aim of the bench study was to evaluate if the ventilator was able to deliver mouthpiece ventilation without alarms and/or autotriggering or ineffective triggering. The 6 most recent life-support ventilators available in France for home ventilation were tested as described.[11] Each ventilator was connected via its standard circuit to a chamber Michigan test lung (MII VentAid TTL, Michigan Instruments, Grand Rapids, Michigan) (Fig. 1). As the PB560 (LX010101/AL020002, Covidien, Mansfield, Massachusetts), Trilogy 100 (13.0, Philips Respironics, Murrysville, Pennsylvania), and VS III (1.12, ResMed, Saint-Priest, France) ventilators have specific mouthpiece ventilation software, all three were also tested with this configuration.

For each ventilator, a pediatric and an adult profile of recorded subjects were tested. The pediatric profile was issued from the record of a 4-year-old patient with spinal muscular atrophy, having a spontaneous tidal volume (V_T) of 10 mL/kg, a compliance of 0.038 L/cm H_2O, and an airway resistance of 52 cm H_2O/L.s. We used a Pneuflo airway resistor Rp50 (Flow Michigan Instruments, Grand Rapids, Michigan), which is characterized by a nonlinear parabolic pressure flow relationship, to create a similar airway resistance. Rp50 introduces a 6.8 cm H_2O pressure

drop for a 0.25 L/s flow and a 27.2 cm H_2O pressure drop for a 0.5 L/s flow. This pediatric profile was tested with 2 volume control settings with V_T 250 mL (12.5 mL/kg) and 500 mL (25 mL/kg), pressure support mode with an inspiratory pressure of 15 cm H_2O, and pressure control mode with an inspiratory pressure of 15 cm H_2O. The adult profile was derived from the record of a 17-year-old patient with Duchenne muscular dystrophy, having a spontaneous V_T of 5 mL/kg, a compliance of 0.024 L/cm H_2O, and an airway resistance of 17 cm H_2O/L.s. We used a Pneuflo airway resistor Rp20, which introduced a 4.4 cm H_2O pressure drop for a 0.5 L/s flow and a 17.6 cm H_2O pressure drop for a 1.0 L/s flow. This adult profile was also tested with 2 volume control settings with V_T 500 mL (12.5 mL/kg) and 1000 mL (25 mL/kg), a pressure support mode with an inspiratory pressure of 20 cm H_2O, and pressure control with an inspiratory pressure of 20 cm H_2O. The backup rate of each ventilator was set at the lowest value, and all of the ventilator alarms were set to maximum. The inspiratory triggers were set at the most sensitive value without autotriggering.

Each ventilator was tested with 3 mouthpieces, a large rigid plastic mouthpiece and a small mouthpiece with or without a filter (Fig. 2). As expected, the resistance was the highest in the small mouthpiece with a filter and the lowest in the large mouthpiece without a filter. (See the supplementary materials at http://www.rcjournal.com.)

For each condition, the following parameters were computed from each pressure and flow trace: trigger time delay (time delay from the onset of inspiratory effort to the

moment during which the airway pressure exceeds the expiratory pressure [ΔT]) and trigger pressure (maximum negative pressure deflection from baseline [ΔP]).[11-13] Signals were digitized at 200 Hz using an analog/digital system (MP100, Biopac Systems, Goleta, California) and recorded on a microcomputer for further analysis.

Concerning the quality of the inspiratory trigger, we used the following criteria for the pediatric profiles only. The inspiratory trigger was considered to be appropriate if $\Delta T \leq 100$ ms and $\Delta P \leq 1$ cm H_2O, acceptable if $\Delta T \leq 150$ ms and $\Delta P \leq 1.5$ cm H_2O, and inappropriate if $\Delta T \geq 150$ ms or $\Delta P > 1.5$ cm H_2O or in case of the non-detection of the inspiratory effort by the ventilator or autotriggering.[11]

Statistical Analysis

Data are given as mean ± SD for quantitative data. Qualitative conditions were compared with a chi-square test. A P value $< .05$ was considered to be significant.

Results

Survey

Completed questionnaires were collected from 209 neuromuscular subjects. Only 11 subjects refused to answer the questionnaire. Thirty of the 209 (14%) subjects used mouthpiece ventilation. Their mean age was 33 ± 11 years, and they had been using NIV for 12 ± 7 years (Table 1). Fifteen subjects used NIV for > 20 h/day, and 11 were totally ventilator-dependent. The majority of the subjects used the same ventilator during the night and day (18/25, 72%). The Eole (Saime, Savigny-le-Temple, France) and Legendair (Covidien) ventilators were used by 77% of the subjects, mostly (75%) in the assist/control mode. Twelve subjects used a double-limb ventilator circuit, 11 subjects used a single-limb circuit with an expiratory valve, and 7 subjects used a single-limb circuit with an intentional leak. The ease of use (or convenience) of the mouthpiece circuit was rated by the subjects as 7 ± 3 on a visual analog scale (0: bad; 10: very good), with 6 subjects having no opinion. The ease of use of the mouthpiece by the subjects was also rated as 7 ± 3, with again 6 subjects having no opinion. As expected, the subjects considered it very important (9 ± 2 on a visual analog scale) to have the mouthpiece as close as possible to their mouths. Seven subjects used mouthpiece ventilation exclusively for prolonged continuous periods of time, 4 subjects used it only for short intermittent breaths, and 8 subjects used it for both short and prolonged continuous periods of time, with no response available for 11 subjects. When the subjects were asked about the frequency, they expired into the ventilatory circuit, the mean response was 4 ± 3, on a visual analog scale (0: never; 10: always), with no response from 2 subjects. Eleven subjects

Table 1. Characteristics of Subjects and Mouthpiece Ventilation

Characteristics	Value
Women/men, n	8/22
Age, mean ± SD, y (range)	33 ± 11 (13–64)
Neuromuscular disease, n	
Duchenne muscular dystrophy	14
Fascioscapular muscular dystrophy	3
Congenital myopathy	3
Becker dystrophy	1
Metabolic myopathy	1
Spinal muscular atrophy	7
Other neuromuscular disease	1
Subjects using a wheelchair, n	29
Subjects with a ventilator on wheelchair, n	21
Duration of NIV, mean ± SD, y (range)	12 ± 7 (2–28)
Daily duration of NIV, mean ± SD, h (range)	19 ± 6 (8–24)
Subjects using NIV > 20 h/day, n	15
Subjects using NIV 24 h/day, n	11
Subjects using ventilator equipment, n	
Same ventilator night and day	18
Different ventilator night and day	7
Not specified	5
Subjects using ventilators with mouthpiece ventilation, n	
Eole	13
Legendair	10
PB560	2
VS Ultra*	2
Harmony†	1
Not specified	2
Subjects using ventilatory mode during mouthpiece ventilation, n	
Assist mode	3
Control mode	4
Assist/control mode	21
Not specified	2
Fixation of mouthpiece, n	
On the patient	10
On the wheelchair	6
On a stand	6
Not specified	8
Use of triggering during mouthpiece ventilation, n	11
Comfort with mouthpiece ventilation triggering/ 10, mean ± SD (range)	7.4 ± 3.7 (1–10)

* ResMed
† Philips Respironics
NIV = noninvasive ventilation

used the inspiratory trigger during mouthpiece ventilation with a visual analog scale comfort reported at a mean value of 7 ± 4 (Table 1). When the subjects were asked if they were interested in increasing the delivered V_T on demand (eg, for airway clearance), 17 subjects were interested, with a mean visual analog scale interest reported at 6 ± 3.
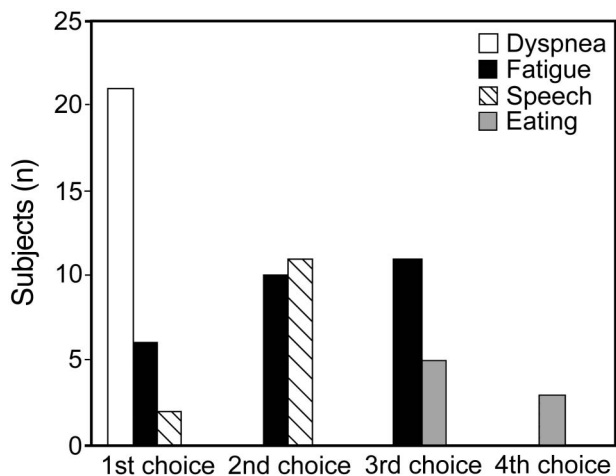
Fig. 3. Subject-reported benefits of mouthpiece ventilation ranked by priority.

Figure 3 shows the subjects' self-estimated benefits from mouthpiece ventilation. The reduction in dyspnea was rated as the most important benefit by 22 subjects (73% of subjects), followed by reduction in fatigue (93%). Facilitation of speech (43% of subjects) and eating (27%) were rated as the third and fourth most important benefits, respectively. Other reported advantages were better vision (23 subjects), reduction in skin injury (19 subjects), facilitation of swallowing (11 subjects), and a decrease in aerophagia (7 subjects). One subject reported that his tracheostomy could be avoided due to mouthpiece ventilation.

Bench Study

The qualitative and partial quantitative performance data on the 3 ventilators with no mouthpiece ventilation software and the 3 ventilators with mouthpiece ventilation software are given in Tables 2 and 3, respectively. (See the supplementary materials at http://www.rcjournal.com.) The main results are that none of the 6 ventilators tested was able to deliver mouthpiece ventilation without alarms and/or ineffective triggering or autotriggering in the different conditions (135 of 198 conditions). For both the pediatric and adult profiles, alarms and/or ineffective triggering or autotriggering was less common with the small mouthpiece and a filter compared to the large mouthpiece without a filter ($P < .001$ for both profiles, $P = .008$ for the pediatric profile, and $P = .039$ for the adult profile). The occurrence of alarms and/or ineffective or autotriggering was not related to the subject profile ($P = .99$), the ventilatory mode ($P = .69$), or the type of ventilator ($P = .95$). However, alarms occurred less often with the VS III ventilator compared to the other devices. Of note, even if the Trilogy mouthpiece ventilator is not designed to trigger

like a classical ventilator, alarms were observed in only 2 of the 18 conditions when a whisper valve was added.

Of the 20 pediatric conditions that were not associated with an alarm and/or ineffective triggering or autotriggering, the trigger was appropriate in only 2 conditions (see Tables 2 and 3). The best triggers were observed with the large or small mouthpiece and a filter. The pediatric triggers were too long with the Elisée ventilator (ResMed) and were associated with autotriggering and/or ineffective triggering for the PB560, Trilogy 100, and Vivo 50 (Breas Medical, Saint-Priest, France) ventilators.

Discussion

This study shows that mouthpiece ventilation is used in highly ventilator-dependent subjects who claim an important subjective clinical benefit from this type of ventilation with regard to a decrease in dyspnea and fatigue and an improvement in speech and eating. These data contrast with the high prevalence of alarms and autotriggering or ineffective triggering observed with 6 home life-support ventilators when used with this type of ventilation.

Survey

The results of our survey confirm that mouthpiece ventilation is effective in postponing invasive ventilation by tracheostomy. Indeed, the majority of the subjects were highly ventilator-dependent, with approximately one third of the subjects being totally ventilator-dependent and half of the subjects requiring ventilatory assistance for > 20 h/day. It is probable that at least some of these subjects would have been tracheotomized in the absence of mouthpiece ventilation.[6,14] This medical benefit is associated with a subjective benefit reported by the subjects. Indeed, all the subjects reported that mouthpiece ventilation was associated with important improvements in dyspnea and fatigue and facilitation of speech and eating, as reported by others.[5,8,14]

Different ventilator circuits were used by the subjects. The use of a double-limb circuit is expected to be associated with fewer alarms if the patient exhales into the expiratory line due to the maintenance of a sufficient minimum pressure to avoid low-pressure alarms. However, our survey revealed that 11 subjects used a single-limb ventilator circuit with an expiratory valve, and 7 subjects used a single-limb circuit with an intentional leak. Regarding exhalation, the subjects' responses were highly variable, with some subjects always exhaling and others never exhaling into the expiratory limb in a double circuit.

The lack of use of the trigger is explained by extreme respiratory muscle weakness. Indeed, subjects with advanced neuromuscular disease are too weak to trigger the ventilator. Only half of the subjects used the trigger during

Table 2. Qualitative and Partial Quantitative Functioning of the 3 Ventilators Without Mouthpiece Ventilation Software With 3 Different Mouthpieces and 2 Patient Profiles (Child and Adult) in the Bench Test

Ventilator	Profile	Ventilatory settings	Alarm response to setting		
			Large mouthpiece without a filter	Large mouthpiece with a filter	Small mouthpiece with a filter
Elisée 150	Child	VC-CMV, 250 mL	LP alarm + open circuit	LP alarm + open circuit	LP alarm + open circuit
		VC-CMV, 500 mL	LP alarm + open circuit	ΔT 0.53 s, ΔP -0.67 cm H_2O	ΔT 0.57 s, ΔP -0.60 cm H_2O
		VC-IMV with PS, 15 cm H_2O	LP alarm + open circuit	LP alarm + open circuit	ΔT 0.55 s, ΔP -0.91 cm H_2O
		PS with security V_T, 15 cm H_2O	LP alarm + open circuit	LP alarm + open circuit	ΔT 0.34 s, ΔP -0.88 cm H_2O
	Adult	VC-CMV, 500 mL	LP alarm + open circuit	ΔT 0.21 s, ΔP -1.33 cm H_2O	ΔT 0.20 s, ΔP -1.13 cm H_2O
		VC-CMV, 1000 mL	HP alarm	HP alarm	HP alarm
		VC-IMV with PS, 20 cm H_2O	LP alarm + open circuit	ΔT 0.19 s, ΔP -1.44 cm H_2O	ΔT 0.18 s, ΔP -1.31 cm H_2O
		PS with security V_T, 20 cm H_2O	LP alarm + open circuit	ΔT 0.19 s, ΔP -1.47 cm H_2O	ΔT 0.18 s, ΔP -1.49 cm H_2O
Monnal T50*	Child	VC-CMV, 250 mL	Patient disconnect	Patient disconnect	Patient disconnect
		VC-CMV, 500 mL	Patient disconnect	Patient disconnect	Patient disconnect
		PC-CMV, 15 cm H_2O	Patient disconnect + high MVI	Patient disconnect	ΔT 0.09 s, ΔP -0.23 cm H_2O†
		PS, 15 cm H_2O	Patient disconnect	Patient disconnect	ΔT 0.08 s, ΔP -0.23 cm H_2O†
	Adult	VC-CMV, 500 mL	Patient disconnect	Patient disconnect	Patient disconnect
		VC-CMV, 1000 mL	HP alarm/patient disconnect	HP alarm/patient disconnect	HP alarm/patient disconnect
		PC-CMV, 20 cm H_2O	High inspiratory V_T alarm	Patient disconnect + high MVI	Patient disconnect + high MVI
		PS, 20 cm H_2O	Patient disconnect	Patient disconnect	ΔT 0.09 s, ΔP -0.33 cm H_2O†
Vivo 50	Child	VC-CMV, 250 mL	LP alarm	LP alarm	LP alarm
		VC-CMV, 500 mL	LP alarm	ΔT 0.11 s, ΔP 0.52 cm H_2O + IT	ΔT 0.11 s, ΔP 0.52 cm H_2O + IT
		PC-CMV, 15 cm H_2O	ΔT 0.18 s, ΔP -0.22 cm H_2O + IT	AT + IT	ΔT 0.45 s, ΔP -0.36 cm H_2O + AT + IT
		PS, 15 cm H_2O	100% AT	AT + IT	ΔT 0.38 s, ΔP -0.25 cm H_2O + AT + IT
	Adult	VC-CMV, 500 mL	LP alarm	ΔT 0.14 s, ΔP -0.34 cm H_2O	ΔT 0.28 s, ΔP -0.30 cm H_2O
		VC-CMV, 1000 mL	ΔT 0.14 s, ΔP -0.17 cm H_2O	ΔT 0.17 s, ΔP -0.40 cm H_2O	ΔT 0.21 s, ΔP -0.44 cm H_2O
		PC-CMV, 20 cm H_2O	ΔT 0.11 s, ΔP -0.37 cm H_2O	ΔT 0.12 s, ΔP -0.51 cm H_2O	ΔT 0.11 s, ΔP -0.44 cm H_2O
		PS, 20 cm H_2O	ΔT 0.14 s, ΔP -0.36 cm H_2O	ΔT 0.15 s, ΔP -0.56 cm H_2O	ΔT 0.09 s, ΔP -0.43 cm H_2O†

* Air Liquide Medical Systems, Antony, France
† Trigger quoted as appropriate for pediatrics (see methods)
VC-CMV = volume controlled continuous mandatory ventilation
VC-IMV with PS = volume controlled intermittent mandatory ventilation (VC-IMV) with pressure support (PS)
PC-CMV = pressure controlled continuous mandatory ventilation
V_T = tidal volume
LP = low pressure
HP = high pressure
MVI = inspiratory minute ventilation
AT = autotriggering > 20% of respiratory cycles
IT = ineffective triggering > 20% of respiratory cycles
ΔT = trigger time delay
ΔP = trigger pressure

Table 3. Qualitative and Partial Quantitative Functioning of the 3 Ventilators With Mouthpiece Ventilation Software With 3 Different Mouthpieces and 2 Patient Profiles (Child and Adult) in the Bench Test

Ventilator	Profile	Ventilatory settings	Alarm response to setting		
			Large mouthpiece without a filter	Large mouthpiece with a filter	Small mouthpiece with a filter
PB560	Child	VC-CMV, 250 mL	Patient disconnect	Patient disconnect	Patient disconnect
		VC-CMV, 500 mL	Patient disconnect	Patient disconnect	Patient disconnect
		PC-CMV, 15 cm H_2O	Patient disconnect	ΔT 0.15 s, ΔP −0.16 cm H_2O + AT + IT	ΔT 0.17 s, ΔP −0.17 cm H_2O + AT + IT
		PS, 15 cm H_2O	Patient disconnect	ΔT 0.18 s, ΔP −0.20 cm H_2O + IT	ΔT 0.24 s, ΔP −0.18 cm H_2O + AT + IT
	Adult	VC-CMV, 500 mL	Patient disconnect	Patient disconnect	Patient disconnect
		VC-CMV, 1000 mL	HP alarm/high inspiratory V_T alarm	HP alarm/high inspiratory V_T alarm	HP alarm/high inspiratory V_T alarm
		PC-CMV, 20 cm H_2O	Patient disconnect	Patient disconnect	ΔT 0.27 s, ΔP −0.46 cm H_2O
		PS, 20 cm H_2O	Patient disconnect	Patient disconnect	ΔT 0.28 s, ΔP −0.47 cm H_2O
PB560 adapted for mouthpiece ventilation	Child	VC-CMV, 250 mL	Patient disconnect	Patient disconnect	Patient disconnect
		VC-CMV, 500 mL	Patient disconnect	Patient disconnect	Patient disconnect
		PC-CMV, 15 cm H_2O	Patient disconnect	ΔT 0.16 s, ΔP −0.20 cm H_2O	Patient disconnect
	Adult	VC-CMV, 500 mL	Patient disconnect	ΔT 0.13 s, ΔP −0.34 cm H_2O	Patient disconnect
		VC-CMV, 1000 mL	HP alarm	HP alarm	HP alarm
		PC-CMV, 20 cm H_2O	Patient disconnect	Patient disconnect	Patient disconnect
Trilogy 100	Child	VC-CMV, 250 mL	Low inspiratory pressure	Low inspiratory pressure	Low inspiratory pressure
		VC-CMV, 500 mL	Low inspiratory pressure	Low inspiratory pressure	Low inspiratory pressure
		PC-CMV, 15 cm H_2O	Check circuit	ΔT 0.07 s, ΔP −0.25 cm H_2O + AT	ΔT 0.07 s, ΔP −0.22 cm H_2O + AT
		ST, 15 cm H_2O	Check circuit	ΔT 0.07 s, ΔP −0.24 cm H_2O + AT + IT	ΔT 0.06 s, ΔP −0.22 cm H_2O + AT + IT
	Adult	VC-CMV, 500 mL	Low inspiratory pressure	Low inspiratory pressure	Low inspiratory pressure
		VC-CMV 1000 mL	Low inspiratory pressure	ΔT 0.10 s, ΔP −0.33 cm H_2O*	ΔT 0.10 s, ΔP −0.29 cm H_2O*
		PC-CMV, 20 cm H_2O	Check circuit	Check circuit	ΔT 0.10 s, ΔP −0.36 cm H_2O*
		ST, 20 cm H_2O	Check circuit	Check circuit	ΔT 0.10 s, ΔP −0.34 cm H_2O*
Trilogy 100 for mouthpiece ventilation	Child	VC-CMV, 250 mL	Leak/disconnect	Leak/disconnect	Leak/disconnect
		VC-CMV, 500 mL	Leak/disconnect	Leak/disconnect	Leak/disconnect
		PC-CMV, 15 cm H_2O	Leak/disconnect	Leak/disconnect	Leak/disconnect
	Adult	VC-CMV, 500 mL	Leak/disconnect	Leak/disconnect	Leak/disconnect
		VC-CMV, 1000 mL	Leak/disconnect	Leak/disconnect	Leak/disconnect
		PC-CMV, 20 cm H_2O	Leak/disconnect	Leak/disconnect	Leak/disconnect
Trilogy 100 for mouthpiece ventilation + whisper valve	Child	VC-CMV, 250 mL	ΔT 0.21 s, ΔP −0.22 cm H_2O + IT	100% IT	100% IT
		VC-CMV, 500 mL	ΔT 0.21 s, ΔP −0.22 cm H_2O	ΔT 0.23 s, ΔP −0.33 cm H_2O	ΔT 0.20 s, ΔP −0.26 cm H_2O
		PC-CMV, 15 cm H_2O	ΔT 0.20 s, ΔP −0.27 cm H_2O	ΔT 0.22 s, ΔP −0.39 cm H_2O	ΔT 0.21 s, ΔP −0.33 cm H_2O
	Adult	VC-CMV, 500 mL	ΔT 0.21 s, ΔP −0.68 cm H_2O	ΔT 0.21 s, ΔP −1.02 cm H_2O	ΔT 0.21 s, ΔP −0.90 cm H_2O
		VC-CMV, 1000 mL	ΔT 0.17 s, ΔP −0.64 cm H_2O	ΔT 0.19 s, ΔP −0.89 cm H_2O	ΔT 0.18 s, ΔP −0.79 cm H_2O
		PC-CMV, 20 cm H_2O	ΔT 0.17 s, ΔP −0.63 cm H_2O	ΔT 0.18 s, ΔP −0.89 cm H_2O	ΔT 0.18 s, ΔP −0.80 cm H_2O
VS III for mouthpiece ventilation	Child	VC-CMV, 250 mL	LP alarm	LP alarm	LP alarm
		VC-CMV, 500 mL	LP alarm	ΔT 0.61 s, ΔP −1.71 cm H_2O	ΔT 0.72 s, ΔP −1.77 cm H_2O
		PC-CMV, 15 cm H_2O	Connect circuit	ΔT 0.61 s, ΔP −1.63 cm H_2O	ΔT 0.61 s, ΔP −1.76 cm H_2O
		PS with security V_T over 3 breaths, 15 cm H_2O	ΔT 0.59 s, ΔP −1.45 cm H_2O	ΔT 0.41 s, ΔP −1.39 cm H_2O	ΔT 0.59 s, ΔP −1.55 cm H_2O
	Adult	VC-CMV, 500 mL	ΔT 0.11 s, ΔP −0.78 cm H_2O	ΔT 0.17 s, ΔP −1.40 cm H_2O	ΔT 0.11 s, ΔP −0.84 cm H_2O
		VC-CMV, 1000 mL	HP alarm	HP alarm	HP alarm
		PC-CMV, 20 cm H_2O	ΔT 0.17 s, ΔP −1.43 cm H_2O	ΔT 0.18 s, ΔP −1.77 cm H_2O	ΔT 0.16 s, ΔP −1.36 cm H_2O
		PS with security V_T over 3 breaths, 20 cm H_2O	ΔT 0.18 s, ΔP −1.54 cm H_2O	ΔT 0.14 s, ΔP −1.18 cm H_2O	ΔT 0.14 s, ΔP −1.11 cm H_2O

* Trigger quoted as appropriate for pediatrics (see methods)
VC-CMV = volume controlled continuous mandatory ventilation
PC-CMV = pressure controlled continuous mandatory ventilation
PS = pressure support
ST = spontaneous/timed ventilation
V_T = tidal volume
HP = high pressure
LP = low pressure
ΔT = trigger time delay
ΔP = trigger pressure
IT = ineffective triggering > 20% of respiratory cycles
AT = autotriggering > 20% of respiratory cycles

mouthpiece ventilation with an acceptable level of comfort. This contrasts with the results of the bench study, which showed a poor quality of the inspiratory trigger in most of the conditions. As all of the subjects had very advanced respiratory muscle weakness, it is probable that they triggered the ventilator by means of a sip. Indeed, the negative pressure generated by a sip is much higher than that generated by a maximum static inspiratory pressure.[15] However, after a sip maneuver, a patient with advanced neuromuscular disease will not be able to generate any inspiratory effort.

Bench Study

The bench study showed the high prevalence of alarms. Logically, open-circuit mouthpiece ventilation can be performed when sufficient peak inspiratory flow is used to create enough back pressure due to mouthpiece resistance to prevent low-pressure alarms in an open-circuit system.[9] As expected, the small mouthpiece with a filter had the highest resistance, which explained the lower occurrence of low-pressure or disconnecting alarms.

We have no explanation for the differences observed between the 6 ventilators. Detailed information on the functioning and performance of home ventilators is generally not available. Indeed, the software with most ventilators is a classical example of a black box. Under United States and European administration regulations, marketing approval for a home ventilator device does not seem to require detailed descriptions of the current algorithms that determine the response of the ventilator to changes in respiratory system mechanics and leaks. In the absence of detailed information, it is thus impossible to explain the differences observed between the ventilators but also between the different ventilatory modes and settings for a particular ventilator. Within the same context, the algorithms that determine the low-pressure or disconnecting alarms vary according to each manufacturer, which precludes any comparison between different ventilators.

Interestingly, the Trilogy mouthpiece ventilator and whisper valve were associated with a reduction in alarms compared to the other configurations available with this ventilator. It should be emphasized that the "kiss" trigger available on the Trilogy mouthpiece ventilator is not comparable to a classical ventilator, as it is based on a flow signal technology that detects an alteration in the flow-by generated by the ventilator due to any reason, such as a partial obstruction via the mouth connection. In our bench test, in which we used a valve to stop the flow-by with variable velocity, preliminary results showed, first of all, that a complete interruption of the flow-by was not necessarily associated with a trigger detection but that a minimum speed of variation of the flow-by (defined as the variation of flow over time) seemed necessary to the trigger detection and the delivery of V_T. After several tests, we observed a threshold of the flow-by of 0.13 L/s^2 necessary for the trigger detection (receiver operating characteristic curve: specificity 96%, sensitivity 87%, area under the curve 0.970, $P < .05$).

Finally, it is important to emphasize the discrepancy between the alarm and inoperable settings observed in the bench test and the subjective satisfaction of the majority of the subjects with their actual mouthpiece ventilation. We do not have a clear explanation for this point, but it is possible that the subjects (and their caregivers) found some way to adapt with the excessive alarms or ventilator dysfunction.

In conclusion, this study shows that mouthpiece ventilation can be a viable treatment option for some subjects with neuromuscular disease and that it is used mainly by highly ventilator-dependent subjects who recognize an important subjective benefit of this type of ventilation. However, there are remarkable technical problems in setting up mouthpiece ventilation. A wider use will require that the equipment be made easier to set up and use properly. Indeed, together with the improvement of ventilator performance for mouthpiece ventilation, there is room for improvement in practical equipment such as mouthpieces and their fixation system and the ergonomy of the ventilators.

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