Mouthpiece ventilation in neuromuscular disorders

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Review article

Mouthpiece ventilation in neuromuscular disorders: Narrative review of technical issues important for clinical success

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ABSTRACT

In neuromuscular disorders (NMDs), nocturnal non-invasive ventilation (NIV) via a nasal mask is offered when hypercapnic respiratory failure occurs. With disease progression, nocturnal NIV needs to be extended into the daytime. Mouthpiece ventilation (MPV) is an option for daytime NIV. MPV represents a difficult task for home ventilators due to rapidly changing load conditions resulting from intermittent connections and disconnections from MPV circuit. The 252nd ENMC International Expert Workshop, held March 6th to 8th 2020 in Amsterdam,

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reported general guidelines for management of daytime MPV in NMDs. This report could not present all the detail regarding the technical issues important for clinical success of MPV. Based on the expert workshop discussions and the evidence from existing studies, the current narrative review aims to identify the technical issues of MPV and offers guidance via a decisional algorithm and educational figures providing relevant information that is important for successful implementation of MPV.

1. Introduction

Patients affected by neuromuscular disease (NMD) often develop progressive weakness of the skeletal and respiratory muscles. Progressive weakness of respiratory muscles inevitably leads to hypercapnic respiratory failure, due to the inability of respiratory muscles to maintain adequate spontaneous ventilation. Respiratory failure is characterized by hypoventilation, an increasing carbon dioxide tension (PaCO2) in the blood, initially during sleep, and later during night and day. A peak PaCO2 during sleep measured by transcutaneous sensor (TcCO2) above 49 mmHg is considered as a major criterion to start ventilatory support in NMD patients. [1] Nocturnal ventilatory support is generally offered via a nasal mask [2], but one third of patient with NMDs require oronasal masks [3]. Clear indicators with regards to the progression of nocturnal ventilation into the daytime are not available. However, when the need to extend nocturnal ventilation into the daytime ventilation is identified, mouthpiece ventilation (MPV) is considered as an essential option for daytime ventilatory support in NMDs [4,5].

The 252nd ENMC International Workshop, held March 6th to 8th 2020 in Amsterdam, the Netherlands, developed best practice guidelines for initiation and management of MPV [6]. Twenty-two international experts from Belgium, UK, France, Portugal, Norway, Canada, USA, Spain, Italy, the Netherlands and Switzerland discussed topics including indications, technical issues, conditions for success, monitoring, safety-risk management, effects of MPV and use in different NMD. Early initiation of MPV was discussed and recommended, especially in the rapidly progressive disorders. Important discussions regarding indications and implementation of MPV, patient preparation, choice of material, monitoring and follow-up, and physiological effects were reported [6].

In practice, MPV can represent a difficult problem to achieve for some home ventilators because of the irregular breathing pattern and rapidly changing load conditions resulting from intermittent connections and disconnections from MPV circuit. Recommendations for specific ventilator devices was not made during the Workshop as device availability varies significantly between countries. However, based on the expert discussions during the workshop [6] and the evidence from existing studies, the present narrative review aims to identify the technical issues related to the use of MPV. It offers guidance for successful implementation and management of MPV. The current review also provides original educational figures illustrating MPV technology, and algorithms to facilitate decisions important for clinical treatment success.

2. Historical development of MPV

Home MPV is not recent technique, dating back even to the polio epidemics of the 1950s. Dr John R Bach, who is still to this day an advocate for MPV for efficient continuous noninvasive ventilatory support, was one of the first clinicians to report mouthpiece/lip seal NIV as an alternative to tracheostomy by 257 patients, most of whom were cared for by Goldwater Memorial Hospital from 1968 in USA [7]. Dr Joshua Benditt and Louis Boitano, from Seattle, USA, presented the first technical-oriented study on MPV, aiming at avoiding disconnection alarms [8]. Then in 2006, the team of Dr Doug McKim in Ottawa, Canada, promoted the combination of nasal and oral rather than invasive interfaces for 24 h ventilatory support [9]. At that time, dedicated circuits and interfaces for MPV were not available, so these pioneers had to adapt and customize NIV equipment. Despite excellent results from aforementioned centers in USA and Canada, MPV was not widely used in other centers until 1996 in Brussels, Belgium, where MPV was used systematically for daytime NIV in Duchenne Muscular Dystrophy (DMD) [10]. At that time, non-dedicated material for MPV was used via piston or bellows volume-cycle ventilators and circuits with exhalation valves. Arm supports were locally custom made with success. Later, a Spanish center started with oral interfaces [11], but, despite increasingly positive results, the use of MPV inspired heated debate and the use of a tracheostomy remained the reference for continuous long-term ventilation at home [12]. In 2010, Bach and Gonçalves described the use of MPV in critical care for successful extubation of 155/157 unweanable patients using a NMD-specific extubation protocol to avoid tracheostomy for long-term ventilatory support [13].

A first physiological study conducted by Toussaint et al., in 2008, reported the benefits of 2-h MPV, provided in the middle of the day, in relieving daytime respiratory muscle loading and related daytime dyspnea in adult DMD patients already receiving effective nocturnal NIV [14]. At the beginning of the second millennium, turbine driven ventilators, pressure-cycling software and single circuits without exhalation valves were increasingly used for NIV in hospital and home [15]. A 6-year period without specific studies on MPV highlights the reluctance of international teams to use MPV, but probably also reflects the difficulties without dedicated devices to widely spread its use. Clearly, additional clinical, technical, and physiological studies were mandatory to understanding and enhancing the clinical use of MPV worldwide. More importantly, there was a critical need to develop specific (dedicated) equipment (ventilator modes, circuits, interfaces and support arms) for MPV to invite medical centers novice to MPV to the safe and effective use of MPV.

In 2012, equipment for MPV was initially developed by Philips Respironics (Murrysville, USA), followed by other manufacturers. See Table 1 for manufacturers and devices that offer a dedicated mode. Finally, specific equipment for MPV was commercially available. Ventilators included software avoiding disconnection alarms and sensitive triggers to accommodate respiratory muscle fatigue. Interfaces included silicone-made mouthpieces to avoid dental deformities and equipment for a smart arm support to facilitate the position of the mouthpiece near the patients lips. Fig. 1 shows the manufacturer available mouthpieces that are on the market.

From 2014, an increasing number of studies conducted in Europe reported technical and clinical evidence regarding the dedicated MPV technology [5,16–18]. Interestingly, the first reports in ALS identified patients who were able to use MPV in this challenging patient population [19,20]. In 2016 and 2018, Dr Ogna and coworkers evaluated ventilators and documented the challenge for ventilators, dedicated or not, to adapt to irregular breathing patterns and rapidly changing load conditions resulting from typical intermittent disconnections from MPV circuit [21,22]. The use of MPV in children with NMDs [23] and the preference of NM patients regarding their favorite equipment for MPV [24] were reported in 2020. Based on the expert workshop discussions [6] and the evidence from existing studies, the current report discusses in depth the specific technical aspects of MPV.

3. Indications and contra-indications for mouthpiece ventilation

The expert group concluded that MPV may be applicable to different

Table 1
Manufacturers and devices that offer a dedicated mode. Mode acronyms (in volume and pressure): abbreviations of MPV volume and pressure modes given by the manufacturer; NA: not available; PEEP: positive end-expiratory pressure; VT: tidal volume; Ti: inspiratory time.

Manufacturer		Philips	Resmed	Breas	Löwenstein	Air Liquide
Device		Trilogy 100, (EVO)	Astral 100, (150)	Vivo 45LS, (55, 65)	LM150TD Luisa	EOVE 150
Mode	volume	AC (A/C-VC)	(A)CV	VCV-MPV	MPVv	MPV
	pressure	PC (A/C-PC)	P(A)CV; PS	PCV-MPV	MPVp	MPP
Alarms	apnea	Off (10–60 s)	Off	Off-15-900 s	Off (5sec-30 Min)	NA
	disconnection	Off-15min	On	NA	NA	NA
	Low pressure	1-40 cmH2O	Off	1–50 mbar	4-60 cmH2O	NA
	PEEP	0-25 (3–25) cmH2O	Off	NA	0-25 cmH2O	NA
Settings	VT (mL)	200-2000 (35–2000)	100–2500	300-2000 (2500)	100–3000	300–2500
	Rate (cycles/min)	0-60 (0–80)	Off (2–50)	0-40 (45LS/55)/0–60 (65)	Off	Off (5–60)
	Pressure	0-50 (3–60) cmH20	2-50 cmH2O	0-50 (45LS/55)/0–60 (65)	4-60 cmH2O	5–59 mbar
	Ti (sec)	0.3–5.0 (0.3–5.0)	0.3–5.0	0.3–5.0	0.2–4	0.3–2.5
Trigger	type	Off, kiss, flow	Touch, High, Med, Low, Off	eSync 1-9	Flow, Sens. 1-10	OFF/AUTO/1-5
Circuits	type	Leak Circuit/Exhalation Valve Circuit	Leak Circuit/Exhalation Valve Circuit	Leak Circuit/Exhalation Valve Circuit	Leak Circuit/Exhalation Valve Circuit	Single leak circuit
Batteries	autonomy battery 1 (h)	4 (7.5)	8 (8)	2.5 (45LS) 4 (55/65)	>6	4
	Autonomy battery 2 (h)	4 (7.5)	8 + 8	6.5 (45LS) 8(55/65)	>6	NA
Weight	Kg	5 (6.3)	3.2	2.4 (45LS) 5.4 (55/65)	3.8	1.3
Programs	Number (n)	2 (5)	2 (4)	3	4	4
Indications		>2.5 Kg, MPV >5 years	>5 Kg	>10 kg (45LS/55) >5 kg (65)	VT > 30 mL	>3.5 Kg

Data from manufacturer's manual guides: Philips Trilogy 100/EVO (Philips Respironics, Murrysville, Pennsylvania, USA); Resmed Astral 100/150 (ResMed, Saint-Priest, France); Breas Vivo 45LS/55/65 (BREAS Medical, Mölnlycke, Sweden); Löwenstein LM150TD Luisa (Löwenstein Medical Technology, Hamburg, Germany); Air Liquide Eove 150 (Air Liquide Medical Systems, Antony, France).

conditions and disorders provided that patients are able to attach the mouthpiece and to perform a maximum insufflation capacity greater their vital capacity [6]. The main indication of MPV is the provision of daytime ventilation in patients with chronic progressive NMD [4,10]. Indications also include other disorders such as severe kyphoscoliosis [25], and COPD during mild, moderate [26] and acute respiratory exacerbation [27–29]. Challenging conditions such as macroglossia, learning difficulties, scoliosis, mild dysphagia and the presence of a tracheostomy are not definitive contraindications. All those conditions worth a trial with MPV. See Table 2 for indications, contra-indications and cautions for the use of MPV.

4. Why circuit leak is a natural part of mouthpiece ventilation?

During long-term nocturnal NIV at home, the respiratory needs of patients, the workload of the ventilator, the resistances in the circuit and respiratory tract of stable NMD patients, are not expected to change over time. In stable conditions, unintentional leaks around the nasal or oronasal mask are unwanted and minimized during sleep.

In contrast to ventilation during sleep, leaks during daytime MPV are intentional and are integral to the success of this technology. The 252nd ENMC expert group considered that, understanding why leaks are normal is key to initiating MPV [6]. During the daytime, the active participation of patients in breathing during their activities contrasts with their passive requirements during sleep: patients are awake, sitting in a wheelchair, moving around, eating, drinking, socialising, using computers and smartphones [18]. Patients also use cough augmentation techniques, when needed [30]. During all these activities, patients with NMD prefer to use MPV than nasal or oronasal interfaces. The reason for preferring MPV is related to their **participation** in controlling their breathing during daytime activities and being able to constantly adjust their breathing to the activity they are engaged in [18].

With MPV, patients intentionally generate a succession of disconnections (partial or complete) and reconnections to the ventilator. In other words, patients choose, with every inspiration, the amount of air which they want to inhale, adjusting the seal with their lips on the mouthpiece. An active, complete seal, ensures an almost full tidal volume (±100%) set in the volume-cycled ventilator, while a more relaxed, incomplete seal will ensure either partial or as little as no inhaled tidal volume (from 0 to 99%).

As such variable proportions of inhaled tidal volumes are accepted by patients, ventilator circuits will have much larger and variable leaks than they are designed to cope with. Theoretically, leaks will not cause discomfort because they are decided and controlled by the patients. This statement is true for volume-cycled modes, but not for pressure-cycled modes. In pressure-cycled modes, leaks can cause discomfort due to uncontrollable leak compensation, leading, in turn, to difficult speaking and perilous swallowing during MPV.

5. Why is volume-cycled ventilation preferred to pressure-cycled ventilation for MPV?

The ENMC expert group agreed in using a volume-cycled ventilation as first line mode to initiate MPV [6]. Volume-cycled ventilators can provide both adequate ventilation, cough augmentation and lung volume recruitment, since it permits breath-stacking to the maximum insufflation capacity (MIC). Despite their availability, such ventilators are infrequently used outside of specialized centers in the care of home ventilated patients, probably because patients start with nighttime ventilation in a pressure mode and are used to this [8]. The management of leaks is the major challenge of MPV that differs according to volume-cycled ventilation *versus* pressure-cycled ventilation [21,22].

5.1. Volume-cycled ventilation

Volume-cycled ventilation implies the setting of a tidal volume. Tidal volume is consistent irrespective of changing mechanical conditions such as leaks, resistances in the respiratory system and patient's effort. Breath stacking is possible, there is no leak compensation and it can be used with a positive end-expiratory pressure (PEEP) set at zero [6]. Fig. 2A illustrates the use of a volume-cycled mode for MPV.

Modern home ventilators have a pneumatic system based on a turbine and perform volume-cycled ventilation using an algorithm to adjust the working pressure based on the previous cycles, corresponding to a

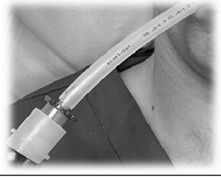
Manufacturer	Description	Reference	illustration
Löwenstein	Mouthpiece for MPV	WM 27646	
Resmed	EasySpeak mouthpiece angled with 17 cm flexitube 22F	21353	
Resmed	EasySpeak mouthpiece angled for 15M connection	21351	
Resmed	EasySpeak mouthpiece angled for 22M/15F connection	21354	
Philips Respironics	Straw Mouthpiece	1105489	
Philips Respironics	Mouthpiece angled	FC06566	

Fig. 1. Manufacturer available mouthpieces that are on the market.
Manufacturers: Philips Respironics, Murrysville, Pennsylvania, USA; ResMed, Saint-Priest, France; BREAS Medical, Mölnlycke, Sweden; Löwenstein, Hamburg, Germany; Air Liquide Medical Systems, Antony, France.

Table 2
Indications, contraindications and caution for using MPV.

Indications	Contraindications	Caution/barriers/risks of failure
Dyspnoea >2.5 on the Borg scale which is relieved by NIV	Inability to perform a lung insufflation capacity which is greater than their vital capacity	Nighttime use
Use of mask ventilation ≥12 h/day	Inability to lip seal around retain the mouthpiece	Fixation of mouthpiece (shoulders/wheelchair) to prevent the patient not been able to reconnect to the mouthpiece
Vital capacity < 30% predicted or dramatic FVC drop (>10%/year)	Inability to control the mouth leaks with MPV	Hypersalivation
Weight loss	Uneven floor when used in the wheelchair	Abdominal distension
Breathlessness when eating	Documented bulbar insufficiency	Orthodontic problems
Breathlessness with talking and/or loss of voice strength	Young age	In case of moderate to severe dysphagia
Weaning from invasive ventilation		Risk for rebreathing when mouthpiece is not detached
Daytime hypercapnia, with normal overnight gas exchange		No arm support available combined with paralysis of the arms

volume-targeted pressure ventilation. This implies a feed-back loop which includes the delivered pressure and the estimated delivered tidal volume (VT), which for its part depends on the actual resistance and compliance of the circuit/patient system. During volume-cycled MPV, this **long feedback loop** results in a slow reactivity of the ventilator to the changing load conditions, with a VT undershoot after reconnection to mouthpiece that stops the leaks [21]. In other words, the actual VT after reconnection is considerably lower than the VT set in the ventilator. This may last a few breathing cycles after firmly reconnecting to the mouthpiece. For this reason, it can be advised to set a higher VT to compensate for VT undershoot which varies between ventilators [21]. The expert group remembered that the old piston/bellows ventilators were excellent for MPV treatment because there was no leak compensation.

Fig. 2A illustrates the use of MPV *via* volume-cycled ventilation. Inspiratory pressure is reduced according to the reduced connectivity to MPV, as suggested during the periods 2 (50% connection to MPV) and 3 (0%). After reconnection to MPV (period 4, 100% reconnection to MPV)), a VT undershoot is seen, which is compensated for by a progressive increase in pressure during the 3 cycles after reconnection to MPV.

In clinical practice, and despite the delivery of lower-than-expected VT with leaks, volume-cycled ventilation allows easy intermittent lip sealing without leak compensation. It is the reason why volume-cycled mode MPV can support even very weak patients. It is intuitive and easy to use, even with imperfect lip sealing such as in Duchenne patients with macroglossia or individuals with weak oral muscles.

5.2. *Pressure-cycled ventilation*

Pressure-cycled ventilation was less preferred for MPV by the members of this working group. We discuss this mode for children who prefer this mode and for those adults who may not have the volume-cycled option. The pressure-cycled mode implies the setting of expiratory pressures (2–4 cmH2O minimum, according to ventilators) and inspiratory pressures. As a result of pressure ventilation, tidal volume is variable and decreases as resistance increases and vice-versa. Breath stacking is not possible as pressure remains constant and does not allow, after the first inhalation, further air intake on the subsequent cycles. There is a leak compensation, PEEP is always present, and rebreathing is possible when they keep the lips around the interface and exhale through the system.

Technologically, the use of pressure-cycled MPV typically results in large volume overshoot during circuit disconnection but minimizes the undershoot in delivered VT at the reconnection to MPV circuit [21]. This can be explained by a **shorter feed-back loop** reestablishing the delivered pressure. This allows adjustments of the ventilator working pressure during the ongoing respiratory cycle. Since the feed-back loop

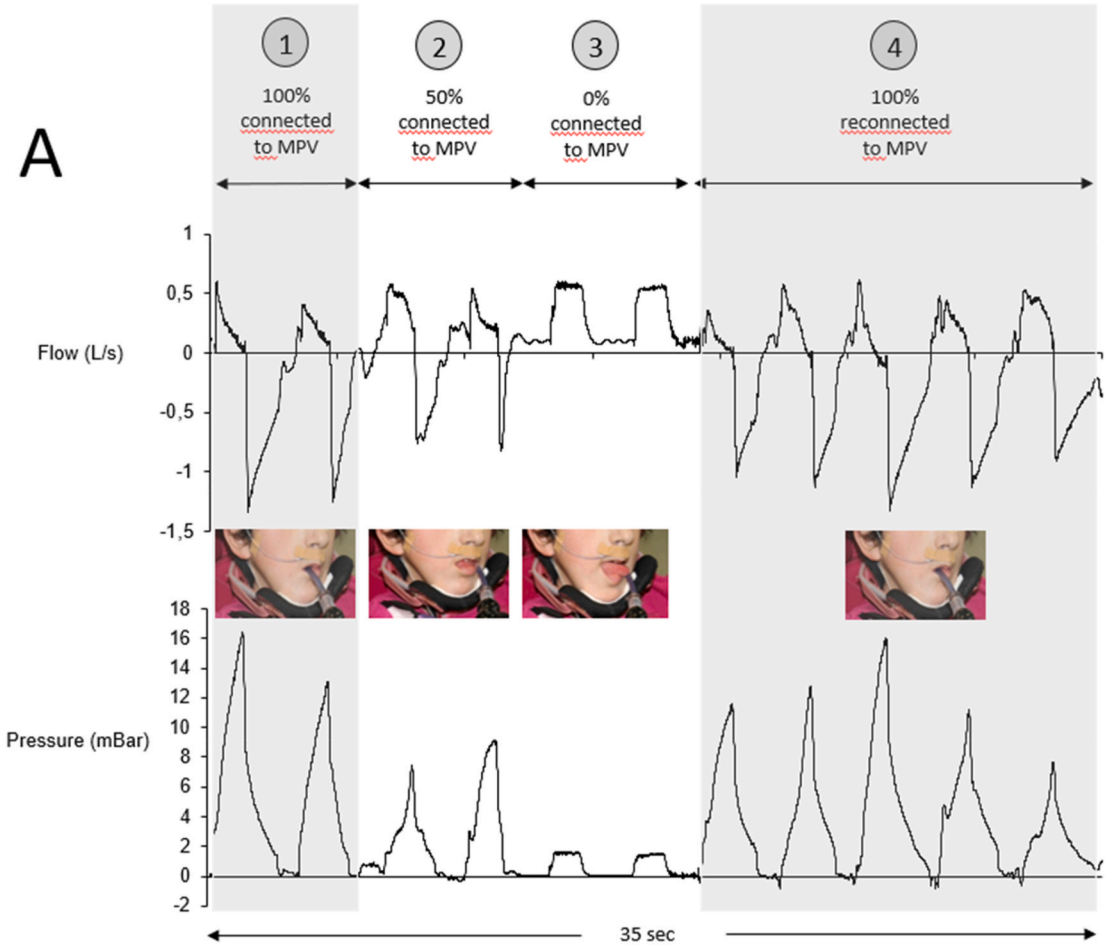


Fig. 2A. MPV via volume-cycled ventilation.
Flow and pressure signals recorded via a heated Fleisch No.2 pneumotachometer (Metabo, Lausanne, Switzerland) during MPV with increasing leakage. MPV via dedicated ACV (volume controlled) mode, Astral 100 (ResMed, Saint-Priest, France).
1, patient seals the lips without leakage; 2, patient seals the lips partially and produces relevant leaks; 3, patient is fully disconnected from MPV and produces massive leaks; 4, patient reconnects to MPV: 3 cycles are needed to recover the preset pressure and volume. We have highlighted in grey the waveforms when the patient is 100% connected to the MPV.

does not include the circuit and the patient, this ventilation mode cannot guarantee a stable VT in the face of changing resistance or compliance of the respiratory system (e.g. in case of airways obstruction due to secretions). This can potentially result in insufficient ventilatory support in highly ventilator-dependent patients. Moreover, in the presence of excessive leaks, those leaks may delay switching to expiration because flow rate is maintained in order to achieve set pressure.

Fig. 2B illustrates the use of MPV *via* pressure-cycled ventilation. Inspiratory pressure is stable regardless of the seal to MPV. As shown during intentional leaks, illustrated by the periods 2 and 3, there is a marked increase in flow to compensate for the leaks. However, the inspiratory pressure is reached immediately upon reconnection to MPV (period 4), suggesting no VT undershoot after reconnection to MPV.

In practice, and by contrast to volume-cycled ventilation, pressure-cycled ventilation requires specific patient's abilities. The expert group advised that patients should be able to easily attach and detach from the mouthpiece with a slight movement of the head (suggested as 5 degrees minimum), and, importantly, to ensure a perfect lip sealing around it with no leaks and, thus, no leak compensation. With pressure-cycled ventilation, the pressure rises rapidly, resulting in a short inspiration time. This may lead to a higher demand for triggered breaths by the patient. To minimize this effect, expiration can be delayed by setting a low expiratory trigger. Table 3 distinguishes the best candidates for a volume-cycled mode from candidates for a pressure-cycled mode.

Using pressure-cycled modes is the intuitive way to set a ventilator because it is extremely common for nigh-time ventilatory support. Centers may just adapt the ventilator from the night-time settings to daytime MPV. The expert group identified using pressure ventilation as a major reason to fail with MPV for the above-described reasons. However, pressure-cycled ventilators can be used as default choice when only bi-level devices, and/or single level devices for 24/7 NIV are available, in areas where there are limited financial resources or when insurance does not cover a second ventilator. Fig. 3 suggests an algorithm regarding the choice of the ventilatory mode.

Pressure-cycled ventilation may be chosen by some children with ventilator free time (spinal muscular atrophy, for example) able to engage and disengage the mouthpiece, who have cognitive skills to use MPV, and who appreciate the rapidly building pressure in their non-compliant lungs and chest wall under any circumstances [6]. It can also be used in children previously using pressure-cycled ventilation before setting volume-cycled ventilation.

6. What is a dedicated MPV mode?

Leaks are normal, intentional, and desired by patients. To cope with the specificities of leak ventilation, industry has developed new software to facilitate the use of MPV. This has been called "dedicated MPV modes", with the aim of providing MPV with all the settings that are needed for the patient to be able to use the technique.

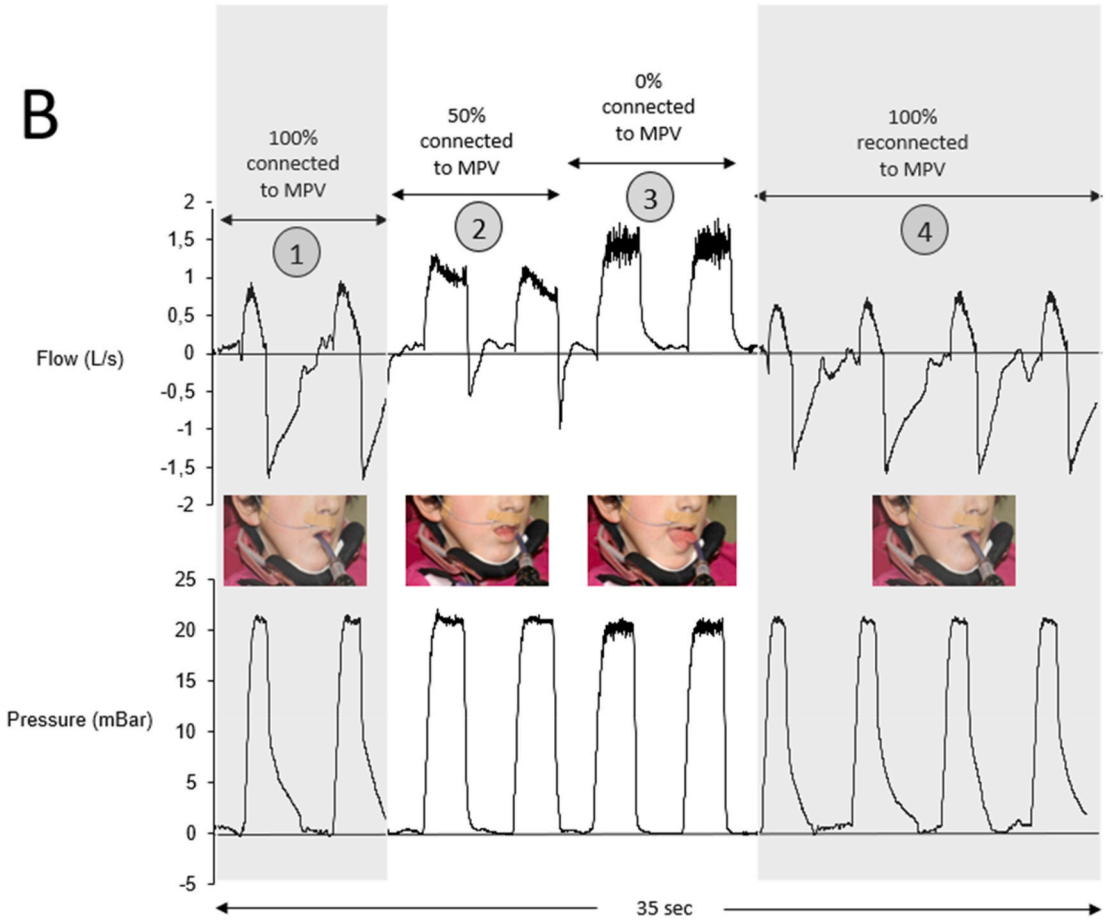


Fig. 2B. MPV via pressure-cycled ventilation.
Flow and pressure signals recorded via a heated Fleisch No.2 pneumotachometer (Metabo, Lausanne, Switzerland) during MPV with increasing leakage. MPV via dedicated PC (pressure controlled) mode, Astral 100 (ResMed, Saint-Priest, France).
1, patient seals the lips without leakage; 2, patient seals the lips partially and produces relevant leaks; 3, patient is fully disconnected from MPV. This produces massive leaks that are compensated as shown by large airflow bursts; 4, patient reconnects to MPV: preset pressure is reached from the first cycle after reconnection to MPV. We have highlighted in grey the waveforms when the patient is 100% connected to the MPV.

Table 3
Patient characteristics required for Volume-cycled *versus* Pressure-cycled mode.

Characteristics of patients	Volume-cycled mode	Pressure-cycled mode
Oral muscle condition	Weak	Preserved
Ability to seal the mouthpiece	Inability	Ability
Ability to engage/disengage from MPV	Inability	Ability
Ventilator dependency	Dependency	No dependency
Ability to use the flow interruption trigger	Fatigue or inability to use it	Ability to use it on the long-term
Possibility to breath-stack	Desired	Not needed
Preference of the type of building pressure	Progressively building pressure	Rapidly building pressure

6.1. The trigger

The new "flow interruption detection" trigger is the most original development in "dedicated" MPV modes. The "flow interruption" trigger does not require an active inhalation or inspiratory effort and, therefore, it is useful for patients with weak respiratory muscles. Instead, patients just need to interrupt the constant flow given by the ventilator, either by pursing the lips around the mouthpiece or by touching the mouthpiece with their cheeks or their tongue[24]. Any flow disruption induces the initiation of an inspiratory cycle, even when the rate is set at zero cycles/minute. Fig. 4 illustrates the attachment and detachment from the mouthpiece to receive such 'on-demand ventilation'.

Patients keeping the mouthpiece between the lips can speak more easily using the volume-cycled compared to a pressure-cycled mode. Indeed, the permanent usage of the mouthpiece in the mouth leads to unwanted triggering. In fact, the trigger is not able to differentiate between intentional or unintentional flow interruptions, which are required for speaking, swallowing or to trigger a breath.

Constantly retaining the mouthpiece, using the dedicated trigger with a backup rate set at zero or at a low value, is therefore challenging when patients wish to speak. When speaking, there is the risk of producing consecutive and unintentional triggers leading to a high respiratory rate. During speech and swallowing, patients do not want air from the ventilator. In practice, the trigger is easier to use in patients able to attach and detach from the mouthpiece as opposed to those retaining the mouthpiece in mouth. Ventilator dependent patients can also feel fatigued as they always need to concentrate on triggering the next inspiration. Under these circumstances, patients may prefer and may require a backup rate [24].

6.2. Respiratory rate at zero

The rate can be set appropriately for patients who may rely on a backup rate due to severe respiratory muscle weakness. Otherwise, it

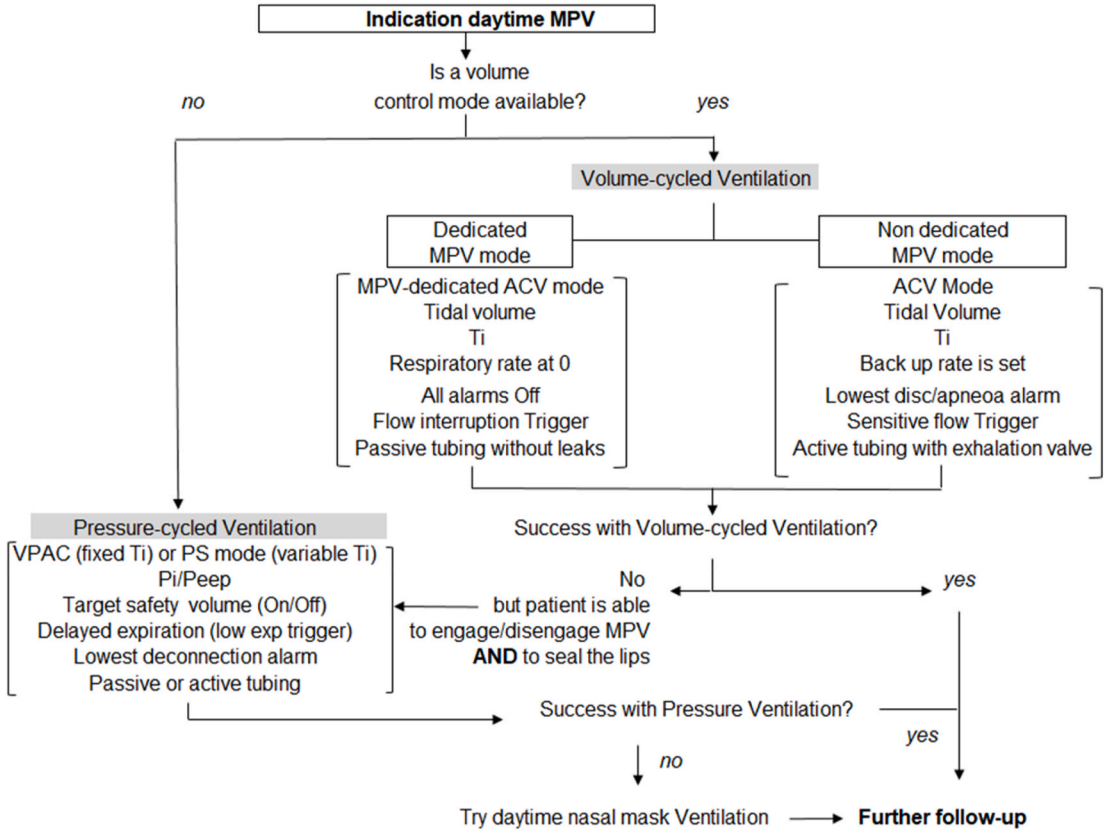


Fig. 3. Algorithm for the choice of the best ventilatory mode for MPV.
MPV: mouthpiece ventilation; MPV dedicated: specific dedicated software and accessories for MPV; ACV: assist-control ventilation; Ti; inspiratory time; flow interruption trigger: see text; passive tubing: tubing without exhalation valve; back up rate: minimal rate set in the device; lowest disc/apnea alarm: alarm activated due to a disconnection or absence of respiratory cycles; active tubing: tubing with exhalation valve; VPAC: assist-control pressure ventilation; PS: pressure support ventilation; Pi: inspiratory pressure; Peep: Positive end-expiratory positive.

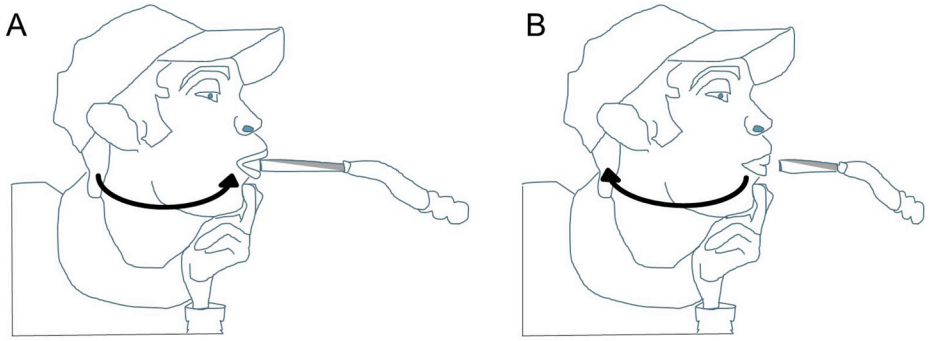


Fig. 4. Illustration of attachment (A) and detachment (B) from MPV.

can be turned "off" or set to zero, depending on the device. Rate at zero is preferred by patients who are able to breath for some periods of spontaneous breathing without dyspnoea [24]. The expert group was not able to determine a minimum ventilator-free breathing time to indicate a backup rate. But, this is probably between 3 and 6 h to use a rate at set to zero [24]. If dyspnoea occurs with a zero rate, then a backup rate needs to be set according to the patients' comfort.

6.3. PEEP set at zero

There is a possibility to set the PEEP at zero or 1 cm H20 in the MPV dedicated modes. In non-dedicated MPV ventilators, EPAP may be mandatory, preventing the suppression of PEEP.

6.4. Apnoea alarm "off"

A relevant advancement provided by the MPV dedicated mode is the possibility to switch the apnoea alarm "off" aiming to supress the normal alarms that would be activated by high leaks and low pressures.

6.5. Combination of passive tubing and volume-cycled ventilation

A passive circuit without a "controlled leak" can be used together with a volume-cycled mode trough MPV dedicated modes. A circuit with an exhalation valve can also be used. The expert group emphasized that patients report having a similar feeling with both circuits. Such similar comfort of ventilation represents a good argument to propose similar circuits for nasal nocturnal ventilation and daytime MPV. By doing so, the ventilator for the night-time can be used in emergency for daytime ventilation, and vice-versa.

6.6. Breath-stacking and lung volume recruitment (LVR)

Breath-stack is possible with volume-cycled ventilation both with active and passive circuits. This is an exclusivity of MPV dedicated software. This characteristic provides effective cough augmentation technique using a mouthpiece. Compliance is diminished because the patient cannot expand the lungs to predicted inspiratory capacity. As the vital capacity (VC) decreases, the largest breath one can take only expands a fraction of the lung volume. Like limb articulations, regular mobilization is required to prevent chest-wall contractures and lung restriction. This can be achieved by providing deep insufflations with breath-stacking.

6.7. Rebreathing alarm

Some devices have a rebreathing alarm. Indeed, no leaks are present in the tubing. This means that there is a theoretical risk for rebreathing. A rebreathing alarm may be activated if the MPV user firmly seals the mouthpiece without creating a single leak within the minute. The expert group reported that rebreathing can occur during the learning process but occurs seldom in trained patients on the longer term, and it is therefore less useful.

6.8. Delayed disconnection alarm

Many ventilators offer the possibility to set a disconnection alarm or not, and to delay its activation for 15 min. The expert group advised to switch this alarm "on" in the very ventilator-dependent patients. This alarm can be set "off" when patients have breathing autonomy and the patient must choose and understand the implications of silencing or delaying this alarm.

6.9. High pressure alarm

According to the manufacturers, there is a possibility to set a high-pressure alarm (55–80 cmH2O, depending on the ventilators). Excessive positive and/or negative pressure can result in pneumothorax [31]. During the discussions, the expert group was not aware of high inspiratory pressures related pneumothorax directly as a result of MPV. The group questioned the usefulness to set highest-pressure alarms, for instance during LVR manoeuvres. Measurement of the maximal inspiratory capacity (MIC) can determine that pressure higher than 55–80 cmH2O is no more effective in achieving that MIC than 50 cmH2O or even less pressure, and perhaps of less risk [6]. Therefore there is little interest to set this high pressure alarm higher than 50 cmH2O.

6.10. Different pre-set programmes

Different programmes (prescriptions) can be pre-set on recent ventilators. The dedicated MPV mode can be one of those programmes. Some ventilators provide multiple pre-set programmes that can be used for nigh-time NIV, daytime NIV, cough assistance, hyper-insufflations (cough augmentation, LVR), and lower respiratory rates for eating, speaking, etc. The night-time mode is settings used by the patient for night-time ventilation and are the settings required to maintain normocapnia via mask ventilation. Often this will be a pre-set pressure support mode. Ventilation during the daytime is often delivered via a volume-cycled mode. Having the ability to switch between mask night-time and mouthpiece daytime settings by pushing a button and confirming the change helps to simplify the changing of settings and limits operational errors.

7. What to do when the MPV dedicated mode is not available or not applicable?

7.1. An alternative volume-cycled ventilator is available

In case MPV dedicated mode is unavailable, the first line choice of mode should be a volume-cycled mode using a circuit with an active expiratory valve. Alarms on the ventilator can be a problem. A correct combination between tidal volume and inspiratory time and the choice of the mouthpiece (in terms of resistance) may avoid disconnection and low-pressure alarms. In case of unexpected alarms, an additional resistance on the circuit, as a filter for example, can avoid the disconnection and/or low pressure alarms [17].

7.2. Only a pressure-cycled ventilator is available

As suggested in Fig. 3, pressure-cycled ventilation can be a default choice for MPV. The conditions of use of bilevel or other pressure-cycled modes has been discussed previously. The EPAP should be set at the lowest available value. In patients with ventilator free time, alarms are useless and should be set at the lowest values. For ventilator-dependent patients, different alarm settings should be considered for safety. In this case, disconnection and apnea alarm as well as a back-up frequency in very weak patients should be set.

8. Conclusions

The main reason identified for MPV failure is the lack of knowledge with regards to the technicalities of MPV. This may result in choosing pressure instead of volume-cycled ventilation in first intention. By contrast to nocturnal ventilation, system leak is part of daytime MPV and one must remember that MPV represents a difficult task to achieve for home ventilators because of rapidly changing load conditions resulting from intermittent connections and disconnections from the MPV circuit. Based on expert panel discussions and on evidence from existing studies, the current narrative review identifies specific technical issues of MPV and offers guidance via a decisional algorithm and educational figures providing relevant information important that is for successful implementation of MPV.

Declaration of competing interest

The authors have no conflict of interest to report in relation to this manuscript.

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