Developing Best Practice Guidelines for Management of Mouthpiece Ventilation in Neuromuscular Disorders

Highlights from the 252nd European Neuromuscular Centre (ENMC) International Workshop (closed session convened in Amsterdam, Netherlands): Developing Best Practice Guidelines for Management of Mouthpiece Ventilation in Neuromuscular Disorders 2020. Large panel of experts on behalf of the ENMC Respiratory Therapy Consortium included: Michelle Chatwin (London UK), Miguel Gonçalves (Porto, Portugal), Jesus Gonzalez-Bermejo (Paris, France), Michel Toussaint (Vlezenbeek, Belgium).

One of the most devastating consequences of neuromuscular disease (NMD) progression is the progressive weakening of muscles of the respiratory system resulting in hypoventilation. This increases the risk of pneumonia and serious comorbidities and ultimately leads to death.[1] In patients with advanced NMDs, ventilatory support is of critical importance both during the day and at night in enabling patient functioning, limiting comorbidities and extending survival. During early stages non-invasive ventilation (NIV) via a mask is sufficient. However, as the disease progresses, bulbar insufficiency worsens and ventilator dependence increases making invasive ventilation methods such as tracheostomy increasingly necessary. These different ventilation methods have notable drawbacks. Quality of life and survival can be improved by NIV[2] but the masks can cause issues such as pressure sores and swallowing difficulties. Tracheostomy on the other hand, can cause pulmonary haemorrhage and loss of speech.[3] A practical alternative is NIV with mouthpiece (open-circuit ventilation [MPV]). This allows daytime support which can be disconnected to allow eating and speaking. The technique is not new and dates back to the polio epidemics in 1953 (Dr John Affeldt)[4] but only became more widely used much later.[5]

MPV has been increasingly used over the past 30 years to increase NIV tolerance and delay or even avoid the need for tracheostomy. It reduces dyspnoea and fatigue and improves patients' speech.

MPV has been increasingly used over the past 30 years as a method to increase NIV tolerance and delay or even avoid the need for tracheostomy.[6] The technique is proven to reduce dyspnoea and fatigue and has also improved speaking capabilities and aids communication.[4] A serious issue in MPV, however, is maintaining the position of the mouthpiece/interface in patients with severe motor impairment. The ENMC Group therefore recommends that patients should undergo a trial of MPV prior to consideration of tracheostomy. They also assert that opting for long-term tracheostomy is a choice only for the patient to make.[7,8] See Table 1 for an overview of ENMC recommendations on MPV use in NMDs.

In NMD patients, day and night time ventilatory support requirements are very different. At night, patients are mostly still, silent and tend to cough and swallow less, so there is decreased need for ventilator autonomy. By day, however, patients are awake, moving, eating and coughing. MPV technology needs to accommodate this with an interface that allows and facilitates these different activities.

MPV is a more convenient alternative to an interface at the patient's face but there have been no randomised studies showing superior survival for this technique over tracheostomy. A recent survey found wide variation in the provision of MPV both within countries and internationally.[7] MPV is usually delivered via a volume-cycled mode of ventilation[9] that delivers a tidal volume of >700ml and ≤1500ml[10,11] without positive expiratory pressure (PEEP), via a mouthpiece or straw supported by a flexible arm/support.[11,12] It is not currently clear which of pressure or volume modes of MPV is superior and whether PEEP has any benefit.[9,10,13,14]

The ENMC Group preference is to start with volume-cycled mode but pressure mode is more applicable in young children. The different interfaces used for MPV are shown in Figure 1. An example MPV system and mouthpiece components are illustrated in Figure 2.

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In MPV, there are multiple different mouthpieces with no consensus on which is optimal or factors leading to choosing a specific type.[15] Treatment centres use either custom-made or commercially produced arm supports; these must be able to move with the patient and maintain their position. Patients should be allowed to try different mouthpieces or a straw and determine what suits them and should also receive anxiety and psychological support from psychology services and advocacy groups.

MPV is a challenge for home ventilators due to irregular breathing, changing load and intermittent disconnection.[12,16,17] More recent home ventilators have a pneumatic system and use an algorithm

Table 1. Overview of European Neuromuscular Centre recommendations for MPV use in patients with neuromuscular disease requiring ventilatory support

Recommendation
All NMD patients should be considered for MPV despite a lack of supportive study evidence for use in specific diseases
MPV can only be used in patients who can maintain a lip seal
Patients with NMD in need of ventilatory support should try MPV prior to consideration of tracheostomy
Patients should be allowed to try different mouthpieces or a straw to determine what suits them
Patients should receive anxiety and psychological support from psychology services and advocacy groups
The ventilator unit should be based on the type of ventilation planned and their advantages/limitations
Patient training of MPV should include teaching patients to assist cough by a single deep breath in pressure mode or 'breath-stacking' in volume mode and how to swallow
Manufacturers need to involve clinicians and patients in ventilator/mask development
MPV usage is largely based on local experience; greater awareness is needed to promote wider usage

Figure 1. Differing interfaces for mouthpiece ventilation

1. Patient using mouthpiece ventilation with a mouthpiece and 2. a straw (Phillips Respironics, Murrysville, PA, US).
Source: Chatwin et al. 2020[7]

to adjust pressure based on previous cycles. This creates a long feedback loop and the ventilator reacts slowly to changing load and tidal volume overshoot during disconnection.[14] This effect is greater during pressure-cycled MPV than with volumetric MPV. It also varies between different ventilator types which need 2–6 cycles to stabilise volume.[13] The choice of ventilator unit should be based on the type of ventilation planned and the advantages/limitations of each one.

MPV is considered the treatment of choice for daytime ventilation and is suitable when ventilatory support is needed >12 hours/day (with or without hypercapnia). MPV is suitable for those with dyspnoea and tachypnoea, however, it can only be used in patients who can maintain a lip seal.[4,18–20]

Timing is also an important issue in MPV implementation. The ENMC Group considered that the optimal timing for MPV initiation should be when:

- Use of mask ventilation is ≥ 12 hours/day.
- There is daytime hypercapnia with nocturnal normocapnia.
- Dyspnoea is relieved by ventilatory support.
- Voice volume needs increasing.
- Cough strength outside the home needed increasing.

MPV at home and in long-term use requires accurate monitoring. A small study of patients using home polygraphy (n=8) found MPV to be effective but most patients could under-ventilate and that effective alarms were vital to alert the patient to the issue.[17] Notable barriers to the provision of MPV at some treatment centres are lack of knowledge and training.[10,11] There is also a lack of protocols for MPV and individualised patient care is required which may not be possible at many treatment centres particularly those unfamiliar with this treatment.[21,22] Patient training is an important aspect of MPV usage; it should include teaching patients to assist cough by a single deep breath in pressure mode or 'breath-stacking' in volume mode.[22,23]

MPV mobility presents technical challenges when used with wheelchairs. The wheelchair must be able to accommodate the equipment and have batteries sufficient to provide life-support for extended periods. Mouthpieces have recently been developed that remain in place when used with patients in a wheel chairs allowing improved mobile ventilation.[24]

MPV has been reported to provide benefits for patients with various different NMDs. These include Duchenne muscular dystrophy for which recent interface developments have enabled more secure ventilation in patients who often cannot hold a mouthpiece.[20] Some patients with amyotrophic lateral sclerosis (ALS) also benefit from MPV in terms of quality of life and reduced hospitalisations.[19] In one study, MPV prolonged tracheostomy-free survival in ALS by 9.5 months but only 19.5% of patients successfully used MPV. MPV has also provided effective ventilation patients with spinal muscular atrophy (SMA) type II. In children with SMA, MPV mouthpieces provide increased ability to speak and participate in school activities and help prevent mid-facial hypoplasia and pectus excavatum.[25] MPV has also shown benefits in patients with congenital myopathies and cervical spine injury.

MPV has shown benefits in patients with Duchenne muscular dystrophy, amyotrophic lateral sclerosis, spinal muscular atrophy, Becker muscular dystrophy, metabolic myopathy, post poliomyelitis and multiple other neuromuscular diseases.

Some small studies report limited MPV use in various other NMDs including: maltase deficiency, myotonic dystrophy limb girdle muscular dystrophy, fascioscapular muscular dystrophy, congenital myopathy, Becker muscular dystrophy, metabolic myopathy, post

poliomyelitis, primary adhalinopathy, congenital dystrophies, Pompe's disease and some other neuromuscular diseases.[9,17,24,26] The ENMC Group concluded that all NMD patients should be considered for MPV regardless of a lack of study evidence for use in specific diseases.[7]

In NMDs, swallowing capability can be can be lost or impaired but this function can be improved during NIV and reduces swallowing fragmentation and dyspnoea.[27] This was highlighted by a study of 10 patients with NMDs showing that MPV improved swallowing compared with spontaneous breathing.[28] Patients receiving MPV should be trained in how to swallow and MPV settings should be adjusted differently to allow eating vs normal breathing. The ENMC Group also recommends that MPV should not be taken away during critical care in patients who use it long-term.

In patients receiving MPV, exercise such as walking can be challenging due the need for a portable respirator. This issue has been partly addressed by some MPV systems allowing physical activity and some have been shown to increase exercise tolerance in those with diaphragm paralysis.[29]

For MPV to be successful, the ENMC Group identified the following conditions/abilities:

- Retention of the mouthpiece.
- Reach the maximal insufflation capacity (>vital capacity).
- Understand the advantages and disadvantages of MPV and tracheostomy.

The ENMC Group also noted that in future, manufacturers need to involve clinicians and patients in ventilator/mask development. They should also provide better arm supports and mouth interfaces and should bench test all new systems to ensure they function adequately. They also stressed that additional work is needed to evaluate MPV effects on speech, swallowing and QoL and that a registry of patients receiving MPV should be established. Improved patient education, MPV passports and social networks could also improve the MPV experience and patient support. Finally, at present MPV usage appears to be mainly based on local experience at particular treatment centres. Greater awareness of and familiarity with the technique among healthcare providers would help improve outcomes for many patients with DMDs who need ventilatory support.

MPV usage appears to be based on local experience. Greater awareness and use of the technique would help improve outcomes for many patients with DMDs who need ventilatory support.

Figure 2. Example of commercially available open-circuit ventilation MPV system (Vivo 45LS – Breas Medical)

Source: Breas Medical

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