CASE REPORT

Intermittent daytime mouthpiece ventilation successfully augments nocturnal non-invasive ventilation, controlling ventilatory failure and maintaining patient independence

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SUMMARY

A 53-year-old woman with spinal muscular atrophy and a 7-year history of nocturnal non-invasive ventilation (NIV) use via nasal mask and chinstrap was admitted electively. Outpatient review suggested symptomatic hypercapnia and hypoxaemia. Use of her usual NIV resulted in early morning respiratory acidosis due to excess mouth leak, and continuous face mask NIV was instigated while in hospital. Once stabilised, she elected to return to nasal ventilation. At outpatient review, respiratory acidosis reoccurred despite diurnal use of NIV. Using the patient's routine ventilator and a novel mouthpiece and trigger algorithm, intermittent daytime mouthpiece ventilation (MPV) was introduced alongside overnight NIV. Control of respiratory failure was achieved and, vitally, independent living maintained. Intermittent MPV was practicable and effective where the limits of ventilator tolerance had otherwise been reached. MPV may reduce the need for tracheostomy ventilation and this case serves as a reminder of the increasing options routinely available to NIV clinicians.

BACKGROUND

This report describes the use of intermittent mouthpiece ventilation (MPV) via an open circuit, using a recently launched mouthpiece and trigger algorithm. It is now routinely available on a standard ventilator and enabled the delivery of daytime and patient-acceptable ventilatory support in the presence of continued respiratory failure despite using non-invasive ventilation (NIV) and standard interfaces.

The patient had previously used nocturnal NIV for the management of hypoventilation, resulting from respiratory muscle weakness due to teenage-onset spinal muscular atrophy (SMA).[1] A nasal mask with chinstrap had always been used due to patient choice, with mouth leak a long-standing problem unsolved by trials of multiple interfaces, altered pressures and inpatient review. When diurnal ventilation became necessary, MPV proved highly successful, unlike the patient's usual interface, in that it was easily adopted by the patient for daytime use and maintained her quality of life and independence despite clear disease progression.

Many other neuromuscular disorders are known to cause hypoventilation and respiratory failure responsive to NIV,[2] hence this case is felt to illustrate an as yet infrequently used[3] but now readily available and effective method of daytime ventilation. It serves as an important reminder to consider the increasing range of options available, and may be expected to offer similar benefits to other neuromuscular disease patients.

CASE PRESENTATION

A 53-year-old woman with an underlying diagnosis of SMA and nocturnal NIV use of 7 years via a nasal mask with chinstrap was electively admitted for optimisation of ventilation. The patient used a pressure-targeted Trilogy ventilator at home (Philips Respironics, Murrysville, Pennsylvania, USA), in spontaneous-timed mode (S/T) with IPAP 22 cm H_2O, EPAP 4 cm H_2O, RR 12 bpm.

A mechanical cough assist device had been issued to the patient 2 years previously, when peak cough flow was measured at 110 LPM, and was in daily use to facilitate secretion clearance (Clearway, B&D Electro Medical, UK). The patient had been unable to perform reproducible respiratory function tests for several years, with her last measured vital capacity 0.81 L (31% predicted) and forced vital capacity 0.87 L (33% predicted) 6 years before this episode.

A 3-week history of general deterioration was described at presentation, with increased work of breathing, hypoxaemia and morning headaches. Overnight domiciliary oximetry prior to admission, wearing NIV without entrained oxygen, had shown ongoing desaturation (mean SpO_2 86%). History and data downloaded from the ventilator indicated that excess mouth leak was again the cause of the patient's ongoing nocturnal hypoventilation. This had been a recurring problem and had been extensively reviewed, and multiple interfaces and pressure settings used without long-term success.

Although the patient was unable to mobilise and had limited upper limb function, she ordinarily led an active social life, independent in a powered wheelchair, out of the house most days, living alone with family support and carers attending four times daily.

INVESTIGATIONS

On the first night of elective admission, NIV was used with the patient's current settings and interface for baseline evaluation. Arterial blood gas sampling performed the following morning showed

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marked hypercapnic respiratory failure (pCO_2 15.6kPa; pH 7.21; St. HCO_3^- 34.2 mmol/L; BE 12.8 mmol/L).

Continuous use of NIV was instigated, and the patient's nasal mask and chinstrap were changed for a face mask, with a clearly measured improvement in delivered tidal volume and reduction in leak. Pressure support was also increased; following these manoeuvres, a reduction in pCO_2 was seen (table 1).

DIFFERENTIAL DIAGNOSIS

No signs of infective or other reversible illnesses were identified, and poor control of chronic respiratory failure due to inadequate ventilation through mouth leak was recognised to have caused the deterioration. Over the remainder of the hospital stay, the patient stabilised and elected to return to nasal mask ventilation with chinstrap. The decision to return to a nasal mask was made by the patient on the basis of discomfort experienced with face mask ventilation, and on concerns regarding the use of a face mask where the user lives alone and is unable to remove the mask in the event of an emergency. A variety of interfaces, delivered pressures (including lower settings in an attempt to reduce leak), and trial of average volume assured pressure support (AVAPS) ventilation did not lead to any maintained benefit.

At the time of discharge, hypercapnia persisted (table 1); pure volume-targeted ventilation was not considered because this was not expected to offer advantage where the cause of the problem was mouth leak. The patient was therefore advised to use nasal mask NIV with chinstrap for two periods of 2 h each day as well as overnight (settings as table 1). Owing to her physical limitations, and need for carer assistance to use the interface and operate the ventilator, daytime NIV use was possible only at home.

TREATMENT

Outpatient review was arranged with the physiotherapy ventilation team, and 2 weeks after discharge the arterial blood gases revealed that respiratory acidosis had returned (table 2), despite adherence to the daytime regime, which was verified by ventilator download. Furthermore, the patient presented with low mood and reported that home daytime utilisation of the ventilator was difficult to coordinate with carers, and hence had severely restricted her social activities, negatively affecting her quality of life.

She agreed to trial intermittent, on-demand MPV for daytime use, the circuit of which could be fixed to the wheelchair and used by the patient when required and without the assistance of carers. This was delivered using the patient's own Trilogy 100 device, and a positionable circuit support arm with disposable circuit and angled mouthpiece (Philips Respironics, Murrysville, Pennsylvania, USA). The MPV feature was launched in the UK in April 2013, and this was our first use of it for community support.

Dual prescription was enabled, to facilitate use of the dedicated MPV feature (pressure control mode, PC) alongside continued use of nasal mask ventilation (S/T mode) overnight.

OUTCOME AND FOLLOW-UP

A month later, the patient was reviewed again, reporting that MPV was well tolerated and felt to be beneficial. Ventilator interaction data for this period were analysed. Mouthpiece tidal volumes (delivered, Vti) exceeded 1000 mL, versus an approximate average 250 mL with nasal mask NIV at comparable pressure settings. Observation of the patient was consistent with achieving higher volumes by MPV than by nasal interface. The download data showed few instances of excessive leak or asynchrony while using MPV, and appeared to demonstrate the ventilatory effectiveness of MPV PC mode delivered by pressure-targeted device.

Table 2 illustrates arterial blood gas results initially sampled off NIV, and repeated after 30 min of routine MPV during this visit; although the latter sample was venous blood, the pCO_2 of 6.13 kPa is the lowest seen for this patient to date. Venous blood was taken for other necessary clinical reasons and the second blood gas was therefore performed opportunistically. The contemporaneous arterial pCO_2 value is not accurately known and cannot be inferred from the venous value, however, it is unlikely to be higher than the venous measurement. It is recognised that peripheral venous pCO_2 has utility as a screen for ventilatory failure whereby a normal venous value essentially excludes significant hypercapnia.[4] The patient disliked arterial sampling and it was deemed to be not needed clinically by the clinician responsible.

The ventilator download data also revealed that MPV was being used multiple times during the day for periods of up to 30 min, meaning that total active use of the ventilator approximated 16 h per day. The patient reported that the MPV was convenient and she had been able to assimilate the use of daytime ventilation into her normal activities, returning to her previous daytime independence. This continues to be the case 10 months later, and no unplanned hospital admissions have occurred.

Table 1 Selected arterial blood gas measurements performed during the acute hospital admission

Date, Time	pH	pCO_2 (kPa)	pO_2 (kPa)	St. HCO_3^- (mmol/L)	NIV therapy	Clinical status
1-5-14, 06.59	7.21	15.62		34.2	IPAP 22, EPAP 4 S/T mode, RR 12 FiO_2 0.24 Nasal mask NIV with chinstrap	Changed to face mask NIV just prior to sampling
1-5-14, 11.11	7.42	7.87	5.51	33.3	IPAP 30, EPAP 4 S/T mode, RR 14 FiO_2 0.21 Face mask NIV	Mask discomfort and aerophagia
12-5-14, 06.11	7.37	7.48	9.86	28.1	IPAP 30, EPAP 6 S/T mode, RR 14 FiO_2 0.21 Nasal mask NIV with chinstrap	Discharge advice to use nasal mask NIV nocte and 2×2 h/daytime

EPAP, expiratory positive airway pressure; measured in cm H_2O; IPAP, inspiratory positive airway pressure; RR, respiratory rate (breaths per minute); NIV, non-invasive ventilation; S/T, spontaneous-timed mode.

Table 2 Blood gas measurements performed during subsequent outpatient review

Date, Time	pH	pCO_2 (kPa)	pO_2 (kPa)	St. HCO_3^- (mmol/L)	NIV therapy	Clinical status
27-5-14, 13.18	7.31	10.10	7.65	30.2	IPAP 30, EPAP 6 S/T mode, RR 14 FiO_2 0.21 Nasal mask NIV chinstrap nocte, and 2×2 h daytime	Sample off NIV. Trial of mouthpiece ventilation after this: mode MPV PC, IPAP 34, EPAP 0, RR 0, FiO_2 0.21
24-6-14, 11.38	7.32	8.35	9.09	26.9	Nocte as above. Daytime MPV: Mode MPV PC IPAP 34, EPAP 0 RR 0, FiO_2 0.21.	Arterial blood taken off NIV and on room air
24-6-14, 13.11	7.43	6.13	5.13	28.2	30 min MPV use as above (FiO_2 0.21)	*Note: Venous blood taken for other clinical reasons.* Patient reports reduced work of breathing on MPV

EPAP, expiratory positive airway pressure; measured in cm H_2O; IPAP, inspiratory positive airway pressure; MPV, mouthpiece ventilation; NIV, non-invasive ventilation; S/T, spontaneous-timed mode; RR, respiratory rate (breaths per minute).

In this case, daytime MPV appeared to offer an acceptable, practicable and effective option where the limits of ventilator tolerance had otherwise been reached and diurnal respiratory failure was present. The patient reported excessive levels of discomfort with face-mask interfaces, and nasal mask ventilation with chinstrap was seen to offer incomplete control of ventilatory failure and a negative impact on quality of life with daytime use.

DISCUSSION

The use of NIV in SMA is based on case series evidence[1] and larger trials in patients with mixed pathology including neuromuscular disease[2]; improvements have been shown in overnight oximetry[1 2] and transcutaneous carbon dioxide measurement.[2] The recent consensus statement on SMA recommends the use of continuous NIV once sleep-disordered breathing progresses to daytime ventilatory failure.[5]

Positionable circuits and programmable mouthpiece modes of ventilation are now readily available to facilitate continuous ventilation, however, a recent review paper reveals the technique is not yet commonly used.[3] The mode and interface used launched in the UK in April 2013, and the patient's routine ventilator is our standard for dependent neuromuscular patients. Globally, selected centres have reported the use of this method[3]; the data are descriptive and were collected before the development of current technology,[6–9] and in one centre, date back to the 1950s.[6] Recipients were wholly[7–9] or largely[6] patients with neuromuscular disease. Two centres[7 8] offered MPV to users of nocturnal NIV at the onset of diurnal ventilatory failure, as was the case here, and suggested that this may avoid the need for tracheostomy ventilation. Our service manages almost 1000 patients with NIV, and continuous ventilation using mouthpiece interfaces has been deployed previously with success. Locally, this was the first instance of intermittent on-demand mouthpiece ventilatory support, controlled by the patient.

MPV is available on the Trilogy ventilator as both volume and pressure targeted modes, and neither is suggested as first choice by the manufacturer. While most reports of MPV have been with volume modes, their superiority has not been demonstrated in a randomised controlled clinical trial. Volume targeted modes may offer additional options such as breath stacking to enhance cough effectiveness. The patient was initially trialled with pressure targeted ventilation, finding it comfortable and effective, with no adverse events, and so it was continued. As we have gained experience with this mode and circuit, subsequent patients have trialled and used a volume targeted mode to similarly good effect. These findings are consistent with recent bench testing of mouthpiece ventilators,[10] with no clearly superior mode or configuration among devices with dedicated MPV software.

For our patient, the possibility of tracheostomy ventilation as part of routine care had been discussed on occasions prior to this episode. She had indicated that she did not wish to receive this, unless a reversible illness occurred where tracheostomy ventilation would offer a temporary solution. Three observational studies have drawn comparison between patients receiving long-term tracheostomy ventilation and NIV/MPV.[11–13] In each of these studies, the two groups differ in characteristics and treatment regimes; hence, comparison of mortality is not possible. However, it appears that greater respiratory morbidity is seen with tracheostomy use[11–13] and fewer tracheostomy ventilated patients remain in their own home, with many reported to be living 'in hospital' or within a dedicated facility.[11 12] Furthermore, qualitative research among ventilator users with neuromuscular disease has revealed that tracheostomy ventilation is associated with greater difficulties in psychological adjustment than NIV;[14] the importance of patient choice regarding the interface, and the prospect of invasive ventilation, was highlighted in particular.

The increasing ability to offer MPV with a wider range of interfaces may therefore provide an effective means to control daytime ventilatory failure, which may otherwise necessitate methods of ventilation that would limit patient autonomy at best, and pose greater clinical risk at worst.

Learning points

- Mouthpiece ventilation may achieve effective control of hypercapnia where nocturnal non-invasive ventilation is being utilised but diurnal ventilatory failure is present.
- Mouthpiece ventilation may offer practical advantages over mask ventilation for daytime use, and clinical advantages of reduced morbidity versus tracheostomy ventilation.
- It is now possible to deliver intermittent mouthpiece ventilation using readily available circuits, novel trigger algorithms and standard ventilators, adding another option for patients and providers of long-term ventilation.

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