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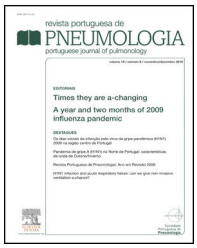

ORIGINAL ARTICLE

Effectiveness and safety of mouthpiece ventilation and nocturnal non-invasive ventilation in patients with kyphoscoliosis: Short and long-term outcomes after an episode of acute respiratory failure

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KEYWORDS
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Abstract
Background: Kyphoscoliosis is a skeletal condition involving the hyperflexion of the thoracic spine. It is characterized by reduced chest wall compliance and impaired respiratory mechanisms leading to progressive hypo-ventilation. We evaluated the effectiveness and the safety of non-invasive ventilation (NIV) in patients after an episode of acute respiratory failure (ARF).
Methods: Eighteen patients with severe kyphoscoliosis who had been hospitalized for an episode of ARF were followed for 4 years. NIV was applied via mouthpiece (MPV) during the daytime and via mask during the night. The primary outcomes were changes in physiological and functional parameters as well as quality of life. Secondary outcomes were considered re-hospitalization and mortality rate after discharge. A set of control subjects was used for comparison.
Results: All patients showed a significant improvement in several clinical, physiological, functional and quality of life parameters. Four of them (22.2%) died during the four year follow-up period. In the uni-variate analysis patients who died had higher cardiac co-morbidity, lower MIP and SNIP, higher $paCO_2$, and oxygen desaturation index at initial admission.
Conclusions: Diurnal MPV associated with nocturnal NIV had significantly improved lung function, clinical outcomes and quality of life. It should be considered as a safe alternative to traditional administering of NIV.

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Introduction

Kyphoscoliosis is a condition characterized by reduced chest wall compliance and impaired respiratory mechanisms leading to progressive respiratory muscle weakness hypoventilation, hypercapnia, and chronic respiratory failure.[1,2] Acute exacerbations are most often due to respiratory tract infections which worsen the respiratory condition. This decline leads to acute respiratory failure (ARF), which may require admission to intensive care (ICU) with either non invasive or invasive mechanical ventilation.[1,3] Non-invasive ventilation (NIV) is the standard long-term treatment because it improves hypoventilation symptoms and reduces sleep fragmentation as well as decreasing hospital, and ICU admissions.[1,3–6] Home NIV originally started during the poliomyelitis epidemic over 50 years ago.[7,8] It continues to be useful because of the increased number of patients who would otherwise remain in the ICU simply because of their ventilator dependence,[4] in this way exposing the patients to iatrogenic complications and decreasing their quality of life, and also increasing costs and it is well-known that the financial burden is an important factor in every health-care system. To avoid these problems, extubation with a safe mode of mechanical ventilation is needed.[1] Pneumonia, respiratory infections and cardiac co-morbidities are the most common reasons for hospitalization.[1,3,9,10] In kyphoscoliotic patients affected by ARF, NIV has a high rate of success (about 75%).[1] It tends to fail in patients with more severe illnesses (patients with sepsis and higher APACHE II scores, and lower Glasgow Coma Score (GCS) and pH values).[1,7,10] Patients may be started on NIV in the ICU or in the High Dependency Unit during ARF where there is minimal independent respiratory function or it may be introduced electively in patients with progressive ventilator insufficiency.[8] Patients with chronic ventilatory insufficiency associated with thoracic restrictive disease derive benefit from long-term ventilation in numerous ways, from improvement in arterial blood gases (ABG) to increased survival rates.[7,8,11] Mouthpiece ventilation (MPV) is a technique that is useful in the treatment of chronic ventilatory failure, particularly in neuro-muscular patients where it has been used for more than half a century.[4,6] MPV is particularly useful when there are problems with conventional masks. The aim of this study is to present the outcomes of kyphoscoliosis patients admitted to a dedicated unit for pulmonary rehabilitation after an episode of ARF, to evaluate the effectiveness and safety of adding MPV to nocturnal NIV in patients with severe ventilatory failure and long term NIV (>12 h ventilation per day),[12] and identify risk factors, readmission and death. NIV failure, number of hospital re-admissions, and mortality at 180-day, 1-year, 2 years and 4-years mortality are examined. Changes in clinical and functional respiratory and quality of life parameters after daytime treatment with MPV plus nasal mask ventilation during the night are presented. Finally, we have compared the outcomes of our cohort with a population of an eighteen control patients treated only with nocturnal NIV.

Table 1 The demographic and anthropometrics characteristics of the MPV patients.

Parameter	Mean ± SD	Range
Age (years)	71 ± 11	44–82
Weight (kg)	74 ± 7	52–81
Height (cm)	169 ± 8	149–175
BMI	26 ± 6	23–33
COBB°	91.6 ± 12.5	82–117
Cardiogenic comorbidities including pulmonary hypertension	7/18 (38.8%)	
Non-cardiac comorbidities	6/18 (33.3%)	

Materials and methods

Eighteen patients were admitted to our institution after an episode of ARF. After discharge each was followed for four years. All had severe kyphoscoliosis with a Cobb angle >80°. Twelve patients had idiopathic kyphoscoliosis, three post-poliomyelitis spinal defects, and three post-tuberculosis complications (Pott' s disease). Their demographic and anthropometric data are shown in Table 1. All patients were new to our center. Each had been admitted to ICU or Emergency Department and treated with invasive or non-invasive ventilation. None had used long-term oxygen therapy or NIV before this study. Inclusion criteria were based on clinical symptoms including morning headache, fatigue, dyspnea, ability to collaborate, and consciousness (GCS ranging between 13 and 15.). Respiratory insufficiency associated with mild/moderate acidosis (pH ranging between 7.35 and 7.30) with persistent hypercapnia ($paCO_2 > 50$ mmHg) was documented in each subject. Absence of systemic inflammation (leukocytes < 10,000/μL), pulmonary parenchymal disease, and absence of recent antibiotic treatment were also necessary for inclusion. Exclusion criteria included impaired consciousness (GSC < 13), initial pH < 7.30, history of bronchial asthma, symptomatic cardiovascular disease, current history of smoking, inability to use MPV and/or refusal to use NIV.

Study design

The study was conducted in a four-bed Respiratory Rehabilitation Unit dedicated to the care of patients requiring non-invasive mechanical ventilation. The study was designed as prospective observational study between May 2010 and September 2014. The study was carried out accordance with the Helsinki Declaration and approved by the local Ethics Committee. All patients provided written informed consent before beginning the study. The study was registered with Chinese Clinical Trials as Chi CTR-OPC 15006661.

Measurements

Before explaining the NIV protocol to each patient, we evaluated dyspnea (Modified Medical Research Council (MMRC) dyspnea scale), Epworth scale (ESS), and SAT-P.[13,14] The patient was asked to assess on a Visual Analog Scale (VAS) his/her satisfaction on 32 life aspects of his/her health status. It provides 32 individual scores and five factors scores ranging from 0 (lowest level of satisfaction) to

100 (highest level of satisfaction).[13,14] Static and dynamic lung volumes[15] and respiratory muscle strength (maximal inspiratory pressure-MIP, maximal expiratory pressure-MEP, sniff nasal inspiratory pressure-SNIP) were measured (Sensormedics VMax 62J Body Plethysmography, CareFusion San Diego, USA).[16] Values of MIP and MEP were obtained at close to residual volume; total lung capacity levels and SNIP values were determined.[14] Ventilatory drive was evaluated by measuring the inspiratory occlusion pressure for the first 100 ms of inspiration (P_{01}) with the patient sitting at rest and breathing room air[17] (Jaeger Masterscreen Pneumo Spirometer). Arterial blood gases were measured when patient awoke (Siemens Rapid Point 405). Blood samples were analyzed for pH, $PaCO_2$, PaO_2, and HCO_{3-}. The CRM of nocturnal events was performed with a portable device 6-channel polygraphy (SOMNO Check Effort, Weinmann V2.05).[18] Respiratory nocturnal episodes (apnea, hypopnea, desaturation) were defined according to the Canadian Thoracic Society guidelines.[19] Trans-cutaneous CO_2 pressure ($tcPCO_2$) was performed using Sen-Tec digital monitor.[20] Finally functional exercise capacity was determined using the six minute walk test (6 MWT).[21]

Non invasive ventilation protocol

NIV was applied via an angled mouthpiece (15 or 22 mm as preferred by the patient) during the daytime and via nasal or oro-nasal mask during the night. The mask that was the best suited to the patient's anatomic characteristics and the ventilator which provided the greatest comfort were selected. All the patients started using assisted controlled ventilation in volume (ACV) during the daytime. The ventilator setting was tidal volume 800–1400 ml respiratory rate 2 rates/min as previously described.[22] The patients gradually became accustomed to the mouthpiece after two to four supervised morning and afternoon training sessions. During the night, patients were treated using assisted controlled ventilation mode in volume (ACV) or if they preferred assisted pressure controlled mode (APCV). The nocturnal ventilatory setting was volume cycled: tidal volume 10–12 ml/kg respiratory rate 16 rates/m′ and pressure-cycled inspiratory pressure 15–20 cmH_2O expiratory pressure 2–3 cmH_2O or lower, as previously described.[5,8] NIV (both nocturnal and diurnal) was implemented using the following ventilators: Breas VIVO 50 or Resmed Elisee 150. Six patients used double limb ventilator circuit; 12 patients used single limb ventilator circuit with expiratory valve. The effectiveness of NIV treatment was evaluated by ABG analysis with the aim of achieving a pH above 7.35, $paCO_2 < 45$ mmHg and $paO_2 > 60$ mmHg. If $SatO_2 > 90\%$ was not achieved, supplemental oxygen was added.

Follow-up

After discharge, the patients had access to the medical staff by telephone and were followed for four years. In the 3rd, 6th, 12th, 24th, 36th, 48th month the patients were evaluated. We examined symptoms, re-admissions, and NIV compliance. Six months after discharge we did complete pulmonary function tests with respiratory functional parameters. Sleep CRM was also repeated.

Outcomes

The primary outcome was the impact of MPV on short clinical, physiological functional parameters and HRQL. Secondary outcomes were considered short-term and long-term hospitalization and mortality after starting of MPV (180–360 days, two years, and four years). Risk factors for mortality were evaluated as well.

Statistical analysis

All data were collected in a databank (Microsoft Access) and analyzed with R-Project software Version 2.13.2. Analysis of covariance was used to test for changes in both parametric and non parametric data at two different time points. Assessment of baseline data between survivors and deceased patients were performed with uni-variate regression analysis. Results are presented as mean values $\pm$ SD for qualitative variables and as frequencies and percentages for qualitative and ordinal variables. p values <0.05 were considered significant.

Results

Ventilation parameters and duration of NIV

All patients were ventilated in ACV mode during the day (tidal volume (VT) 1100 ± 400 ml) and ACV or APCV mode during the night. Initially nocturnal ventilation was ACV mode in six patients; twelve chose APCV mode. During the treatment two patients switched from ACV to APCV mode because of side effects (aerophagia). The settings for ACV mode were a respiratory frequency 16 ± 2 cycles with VT 1000 ± 300 ml. The settings for APCV were: inspiratory pressure (IPAP) 22 ± 4 cmH_2O and expiratory pressure (EPAP) 2 ± 1 cmH_2O. NIV (diurnal plus nocturnal) was used on average 16 ± 3 h daily from discharge to six months. None of the study patients used the ventilator for less than 12 h per day during the follow-up period. NIV usage was determined by a specialist nurse at every outpatient clinic visit. She checked the built-in time counter of the ventilator. Four patients (22.2%) used it less than 14 h; five patients (27.8%) 14–16 h and nine (50%) patients more than 16 h per day.

Effects of NIV

All patients who received MPV and nocturnal NIV showed a significant improvement in several clinical parameters as well in ABG parameters (PaO_2/FiO_2, $PaCO_2$ and pH) and improvements were observed in FVC, FEV_1, MIP, MEP and SNIP, VT, and breathing frequency, P_{01}. 6MWT distance improved, but it did not achieve statistical significance. Polygraphic data also improved significantly as did $tcPCO_2$. We found a discrepancy between apnea-hypopnea index (AHI) and oxygen desaturation index (ODI) because in some patients we observed episodes of apnea in order to no diaphragmatic activity > 10 s without significant desaturation >4%. HRQL and SAT-P scores related quality of sleep ($p < 0.01$); physical well-being ($p < 0.003$); sleep, eating, and leisure ($p < 0.02$); self-confidence ($p < 0.01$); and

Table 2 Lung function, breathing pattern and health-related quality of life measurement in the MPV group at admission and after 6 months treatment.

Variable	Pre-treatment (admission)	Post-treatment(after 6 months)	*p*-value
FVC	1698 ± 493	2216 ± 367	0.02
FVC%	35 ± 6	51 ± 5	0.02
FEV_1	1342 ± 388	1647 ± 391	0.04
FEV_1%	36 ± 3	52 ± 9	0.03
FEV_1/FVC%	80 ± 7	87 ± 3	0.01
TLC	4.390 ± 1045	4600 ± 10.57	0.08
TLC%	68 ± 7	82 ± 8	0.07
VT ml	370 ± 88	428 ± 65	0.03
Breathing frequency	21 ± 2	16 ± 2	0.004
TI/T TOT	0.44 ± 0.03	0.38 ± 0.02	0.05
VT/TI (ml/sec)	377 ± 121	411 ± 89	0.09
P_{01}	0.24 ± 0.11	0.16 ± 0.05	0.03
P_{01} max	3.95 ± 1.93	4.96 ± 1.70	0.06
P_{01}/Pmax	0.08 ± 0.02	0.05 ± 0.02	0.83
P_{01}/MV	0.06 ± 0.03	0.05 ± 0.01	0.78
MIP (cmH_2O)	38 ± 13	46 ± 9	0.04
MEP (cmH_2O)	40 ± 12	45 ± 7	0.08
SNIP	46 ± 8	55 ± 9	0.005
6 MWD (mt.)	272 ± 57	311 ± 68	0.06
paO_2	59 ± 6	66 ± 4	0.06
$paCO_2$	56 ± 6	44 ± 3	0.03
pH	7.35 ± 0.07	7.37 ± 0.01	0.03
HCO_{3-}	32 ± 4	27 ± 4	0.009
PaO_2/FiO_2	265 ± 28	314 ± 18	0.04
Dyspnea (MMRC scale)	3 ± 1	2 ± 1	0.06
EPSS (Epworth Scale)	16 ± 2	8 ± 2	0.004
Apnea-hypopnea index (AHI)	26 ± 18	10 ± 4	0.002
Oxygen desaturation index (ODI)	20 ± 6	13 ± 3	0.001
Mean oxygen saturation	86 ± 2	93 ± 1	0.005
Nadir $SatO_2$%	70 ± 8	88 ± 4	0.002
Average drop	8 ± 2	5 ± 1	0.003
Time desat	47 ± 6	11 ± 4	0.002
T 90%	18 ± 4	1 ± 1	0.002
Tc PCO_2 mean	56 ± 6	46 ± 4	0.003
SAT-P Amount and quality of sleep	39.20 ± 21.11	56.66 ± 20.44	0.01
SAT-P Physical well- being	40.56 ± 27.88	62.68 ± 19.88	0.003
SAT-P Sleep, eating, leisure	45.34 ± 19.57	55.47 ± 16.56	0.02
SAT-P Self-confidence	51.32 ± 26.27	67.21 ± 19.40	0.01
SAT-P Mood	42.16 ± 28.55	61.05 ± 19.88	0.02

paO_2, arterial oxygen pressure; $paCO_2$, carbon dioxide arterial pressure; MIP, maximal inspiratory pressure; MEP, maximal expiratory pressure; SNIP, sniff nasal inspiratory pressure; $P0_1$, inspiratory occlusion pressure for the first 100 ms of inspiration; VT, tidal volume; TI, inspiratory time; AHI, apnea hypopnea index; ODI, oxygen desaturation index; $tcPCO_2$, transcutaneous CO_2 pressure; FVC, forced vital capacity; FEV_1, forced expiratory volume 1 s; TLC, total lung capacity; HCO_{3-}, bicarbonate; MMRC scale, Modified Medical Research Council dyspnea scale; PaO_2/FiO_2, arterial oxygen pressure/fractional inspired oxygen ratio; SAT-P, Satisfaction Profile.

mood ($p < 0.02$) showed statistically significant improvement (Table 2).

Hospital re-admissions and factors related to mortality

All the patients were alive at six months post discharge. One patient with severe pulmonary hypertension was admitted to ICU because of an episode of severe ARF and died nine months after initial discharge. Another patient died within the first year from severe ARF. Two other patients died during follow-up: both were older than 75 years of age and had severe cardiac or cardio-pulmonary (e.g., pulmonary hypertension) pathologies. Both were re-admitted to ICU with severe ARF. Clinical outcomes are presented in Table 3. Eight patients (44.4%) were readmitted during follow-up to hospital. Four of the 18 patients followed died (22.2%). Causes of readmission were mainly due to respiratory infections (42.9%) and severe ARF. Uni-variate analysis of mortality demonstrated positive correlations with cardiac co-morbidity, lower MIP, lower SNIP, higher $paCO_2$, and oxygen desaturation index (ODI) at baseline admission (Table 4).

Table 3 Clinical outcomes of patients treated with MPV.

Patients readmitted to hospital	8 (44.4%)
Number of readmissions	14 (100%)
ICU readmission	4 (28.5%)
Causes of hospital readmission	14
Respiratory infections	6 (42.9%)
Severe respiratory failure	4 (28.5%)
Cardiac failure	2 (14.3%)
Other causes	2 (14.3%)
180 day mortality	1 (5.55%)
1 year mortality	2 (11.1%)
2 year mortality	2 (11.1%)
4 year mortality	4 (22.2%)
Overall mortality	4 (22.2%)

Table 4 Variables associated with mortality in MPV.

Variable	Survivors 14	Deaths 4	*p*-value
Age	69 ± 8	75 ± 6	0.39
Sex (male)	7/14	3/4	0.06
FVC	1734 ± 399	1597 ± 492	0.11
FEV_1	1399 ± 361	1320 ± 394	0.09
TLC	4460 ± 994	4230 ± 1070	0.06
paO_2/FiO_2	269 ± 23	254 ± 32	0.09
$paCO_2$	53 ± 9	62 ± 6	0.05
pH	7.36 ± 0.02	7.35 ± 0.04	0.04
MIP	42 ± 7	34 ± 7	0.04
MEP	40 ± 3	38 ± 4	0.09
SNIP	49 ± 6	42 ± 3	0.05
P_{01}	0.22 ± 0.09	0.24 ± 0.07	0.09
VT	356 ± 64	348 ± 52	0.12
Respiratory frequency	17 ± 2	21 ± 3	0.04
AHI	23 ± 10	26 ± 8	0.08
ODI	36 ± 10	47 ± 9	0.03
Epworth scale	13 ± 1	15 ± 2	0.08
Comorbidities	2	4	0.06
Cardiac comorbidities	1	6	0.01

paO_2, arterial oxygen pressure; $paCO_2$, carbon dioxide arterial pressure; MIP, maximal inspiratory pressure; MEP, maximal expiratory pressure; SNIP, sniff nasal inspiratory pressure; $P0_1$, inspiratory occlusion pressure for the first 100 ms of inspiration; VT, tidal volume; AHI, apnea-hypopnea index; ODI, oxygen desaturation index; $tcPCO_2$, transcutaneous CO_2 pressure; FVC, forced vital capacity; FEV_1, forced expiratory volume 1 s; TLC, total lung capacity.

Table 5 Comparison of the MPV group and the control group.

Variable	MPV group	Control group	*p*-value
Female	11	12	0.28
Male	7	6	0.33
Age	71.4 ± 10.9	69.1 ± 8.8	0.52
Height	163.9 ± 9.2	167.7 ± 4.1	0.19
Weight	74.0 ± 7.3	69.8 ± 4.9	0.19
Comorbidities	9	8	0.11
Cardiac comorb	7	5	0.09
ΔpaO_2	10.4 ± 4.9	8.5 ± 5.4	0.08
$\Delta paCO_2$	−16.0 ± 6.6	−9.1 ± 4.3	0.09
ΔpH	31.2 ± 19.6	17.9 ± 13.6	0.07
ΔHCO_{3-}	−6.6 ± 2.2	−3.5 ± 1.8	0.06
ΔFVC	115.8 ± 486.3	2.2 ± 651.1	0.01
ΔFEV_1	76.2 ±195.7	14.5 ± 211.5	0.03
ΔTLC	188.9 ± 1051.0	26.7 ± 554.0	0.02
ΔMIP	6.7 ± 13.2	3.7 ± 7.7	0.02
ΔMEP	6.4 ± 12.6	5.1 ± 7.3	0.05
ΔSNIP	9.2 ± 8.3	2.9 ± 6.1	0.03
$\Delta P0_1$	−0.1 ± 0.1	−0.1 ± 0.1	0.14
Δ6MWT	19.5 ± 8.4	9.3 ± 7.2	0.04
ΔBR	−1.8 ± 1.8	−2.0 ± 1.5	0.07
ΔEPSS	−6.8 ± 2.1	−5.8 ± 2.2	0.12
ΔAHI	−15.9 ± 13.2	−17.5 ± 10.0	0.19
ΔODI	−22.4 ± 10.5	−18.9 ± 7.5	0.24
ΔMMRC	0.5 ± 0.0	0.5 ± 0.0	0.88
ICU readmissions	28.5%	30.2%	0.07
180 day mortality	5.55%	7.93%	0.03
360 day mortality	11.1%	12.8%	0.03
720 day mortality	22.2%	25.6%	0.05
1440 day mortality	22.2%	23.4%	0.06

paO_2, arterial oxygen pressure; $paCO_2$, carbon dioxide arterial pressure; MIP, maximal inspiratory pressure; MEP, maximal expiratory pressure; SNIP, sniff nasal inspiratory pressure; $P0_1$, inspiratory occlusion pressure for the first 100 ms of inspiration; VT, tidal volume; AHI, apnea-hypopnea index; ODI, oxygen desaturation index; FVC, forced vital capacity; FEV_1, forced expiratory volume 1 s; TLC, total lung capacity; HCO_{3-}, bicarbonate; MMRC scale, Modified Medical Research Council dyspnea scale.

Comparison with a historical group treated with nocturnal NIV

The comparison of the group treated with MPV and nocturnal NIV and historical group treated with nocturnal NIV (plus diurnal NIV only if needed)is reported in Table 5. The control patients suffered from severe kyphoscoliosis previously treated with NIV without adding daytime MPV. Each patient used NIV at least 12 h per day. There was no statistically significant difference in the time NIV was used between the two groups (15.3 ± 4.6 vs 12.2 ± 2.1 $p < 0.09$). These patients were treated with the following home ventilators: Resmed Idea Ultra, Resmed VS III, Resmed Elisee 150,Breas Vivo 50 and Legendair Covidien. Eight of them were treated with APCV and double limb ventilator circuit; ten of these used pressure support ventilation (PSV) with security or guaranteed volume and single limb circuit with expiratory valve. Patients receiving NIV during night time showed an increase in 180–360 and 720 day mortality compared to MPV group. Moreover the MPV group had a significant improvement of some respiratory function parameters after 6 months treatment (FVC, FEV1, TLC, MIP,S NIP, 6MWT).

Discussion and conclusions

Our findings demonstrated improvement in clinical symptoms, blood gases and nocturnal ventilation, sleep related parameters and HRQL scores in our patient cohort. These improvements were accompanied by a significant increase in lung volumes and respiratory muscle function following diurnal ventilation via angled mouthpiece with nocturnal ventilation via mask. Ours is the first study describing management of kyphoscoliotic patients following an episode of ARF using diurnal MPV associated with nocturnal ventilation via a mask. Most previous studies investigated stable patients who were given nocturnal ventilation[4,5,8,9,11,23–27] and demonstrated the effectiveness of nocturnal ventilation in relieving symptoms, improving ABG accompanied by a significant increase in lung volumes and inspiratory muscle function.[5,8,24–28] A previous study reported that patients with ARF receiving 24 h/day non-invasive ventilation for a few weeks significantly improved and returned to baseline condition after about four weeks.[3] Only one study assessed acute kyphoscoliotic patients and their follow-up, considering not only improvements in clinical symptoms, ABG or lung function, but also treatment compliance, hospital and ICU readmission and treatment compliance.[1] Twenty-nine patients were followed for at least 12 months: eight patients were readmitted 19 times.

Five of them died (a 4 years mortality rate of 20.7%). Only pH and 6MWT distance significantly improved after NIV.[1] No improvement was shown in $paCO_2$, paO_2/FiO_2 or lung volumes. NIV in pressure-support mode was used with an IPAP set to obtain VT of 8–10 ml/kg. The authors concluded that NIV improves pulmonary performance in the long term.[1] The mortality rates of kyphoscoliosis patients with long-term domiciliary NIV therapy has been reported to be 9.5% at two years, 20.7% at four years up and 28.1% at five years.[1,29,31] Predictors of mortality were rarely investigated, nor were co-morbidities. Marti et al.[31] suggested that a high level $paCO_2$ (>50 mmHg) at 1 month after starting NIV and the presence of co-morbid conditions are risk factors for mortality in patients with kyphoscoliosis and cardiopulmonary co-morbidity may contribute to higher $paCO_2$ and lower paO_2 level, worsening the prognosis of these patients.[10]

Our data are consistent with these previous studies. Patients who had higher cardiac co-morbidities, lower pH, MIP and SNIP and higher respiratory frequency, $paCO_2$ and ODI at admission tended to have higher mortality. In her study Kirani[32] evaluated 209 patients (30 in MPV) using a questionnaire. Her patients reported that MPV use was correlated with a reduction of dyspnea and fatigue along with greater facility in speaking and eating. One subject thought that his tracheotomy could have been avoided if he had started using MPV earlier. In a case report Ward[33] described a patient with severe diurnal acidosis who refused oro-nasal mask. Standard NIV was not effective. The use of diurnal MPV provided better control of respiratory failure, allowed independent living as well as avoiding tracheotomy.

Our study has some limitations: the number of patients followed by a single center; sleep data were limited; clinical and functional data were only recorded at six month follow-up visit and thus cannot be correlated with mortality or hospital re-admission.

In conclusion, NIV using a combination of mouthpiece and mask in severely kyphoscoliotic patients improves both clinical and functional parameters. As clinicians we can offer more options by adding daytime MPV: it is a practical and safe way to treat ventilatory insufficiency; this modality promotes independent living and provides these patients an enhanced quality of life.

Conflicts of interest

The authors have no conflicts of interest to declare.

Ethical responsibilities

Protection of human and animal subjects. The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Confidentiality of data. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

Right to privacy and informed consent. The authors declare that no patient data appear in this article.

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