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Intermittent abdominal pressure ventilation: feasibility and efficacy in neuromuscular disease. A case report

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Abstract

The standard treatment for patients with neuromuscular respiratory failure is non-invasive ventilation (NIV) as non-invasive ventilation support-setting (NVS). NVS is administered through a nasal or face mask and/or mouthpiece with the potential to cause nasal ulcers, discomfort, and/or aesthetic issues, resulting in poor compliance. We reported the observation of a 45-year-old woman with limb-girdle muscular dystrophy (LGMD), secondary to Dysferlin deficiency, who was on NVS since 2017 for nocturnal hypoventilation. In 2018, despite nocturnal ventilation, due to weight gain and daytime hypoventilation, a nasal mask was introduced. We initiated daytime intermittent abdominal pressure ventilation (IAPV) to mitigate cosmetic problems, improving in pO_2 and decreasing in pCO_2 *versus* baseline (52>84 mmHg, 46>33 mmHg, respectively) at 6 (85 mmHg, 42 mmHg) and 18 months (93 mmHg, 38 mmHg), respectively. IAPV was effective, safe, and well-tolerated in our patients who did not tolerate standard daytime NVS with the known interface.

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Introduction

Non-invasive ventilation support-settings (NVS) are often needed initially at night in patients with neuromuscular disease. However, when the condition worsens, it becomes necessary even during the day [1-3]. Facial masks (nasal, oronasal, nasal pillows, and mouthpieces) are the most used to convey NVS [1]. However, interfaces can cause skin discomfort, ulceration, dryness, and congestion of the airways, negatively affecting the quality of life and gas exchange [1]. In our study, the IAPV was performed by a portable ventilator with an internal battery and the PBAir™ corset as an interface. The IAPV corset is lightweight (Figure 1), comfortable, and, thanks to Velcro fasteners, easier to wear than a face mask. Cyclical inflation of a rubber bladder inside the corset moves the diaphragm upwards. This causes air to enter the lungs via the upper airway as gravity draws the diaphragm back to its resting position.

Case Report

In March 2019, we admitted to our hospital a 45-year-old woman (BMI 30.1 kg/m^2), with limb-girdle muscular dystrophy (LGMD), secondary to Dysferlin deficiency, for persistent daytime desaturation in spontaneous breathing. Since 2017, she had been on night-time NVS with Monnal T50 (Vitalaire, Assago, MI, Italy), compliant for about 8 h/night. Moreover, due to pulmonary infection, she used mechanical insufflation-exsufflation at home. In particular, due to cough weakness, ATOS 70 (Vitalaire, Assago, Italy) was set at insufflation pressure: +45 cmH_2O, inspiratory time: 2.2", exsufflation pressure: -45 cmH_2O, expiratory time: 1.8 and pause: 1.0. Daytime sessions of the lung volume recruitment

by her ventilator in vol-preset tidal volume (TV) 1100 ml, respiratory rate R 4/min and ZEEP (zero end-expiratory pressure) twice a day were assured by nasal pillows interface Swift FX M (Resmed, UK). Last year, she reported increased dyspnea at rest, associated with respiratory exacerbations (2/year). Therefore, the patient increased NVS during the day, for at least 18 hours/day.

Clinical findings

Table 1 shows the results of the arterial blood gas (ABG) in spontaneous respiration, lung function tests, and the night-time polysomnography in NVS at admission.

Night-time NVS was set in assist pressure control ventilation (APCV) mode with VT Target 7 mL/kg and inspiratory positive airway pressure (IPAP): 24/30 cmH_2O, expiratory positive airway pressure (EPAP): 3 cm H_2O, RR 15/min, inspiration/expiration (I:E) rate at 1:1.4 by a Simplus Full-Face Mask (Fisher & Paykel, Milan, Italy). The daytime ventilation parameters were: IPAP 24 cmH_2O, EPAP 3 cmH_2O, RR 12, and I: E 1:1.4 without VT target by nasal pillows interface Swift FX M (Resmed).

Timeline

T0 – February 2019: we introduced IAPV after severe diurnal hypoventilation, during hospitalization.

T1 – after 6 months: first follow-up.

T2 – after 18 months: second follow-up.

Therapeutic intervention

We admitted the patient for respiratory rehabilitation due to daytime hypoxia and hypercapnia in spontaneous breathing, requiring continuous NVS due to weight gain. The patient complained of daytime NVS because of ulcers in her nostrils and refused mouthpiece ventilation as other proposed nasal interfaces, continuing to use the nighttime oronasal mask. Therefore, we used the IAPV as an alternative to daytime NVS. IAPV (LUNA, Dima, Bologna, Italy) was set with IPAP at 60 cmH_2O, rise time at 0.5 sec, and RR at 15 min, and the belt was adapted to her abdominal shape. The patient used IAPV for 10 hours/day for 12 days, without any side effects and it was well tolerated.

Figure 1 shows the woman wearing her corset with the LUNA device.

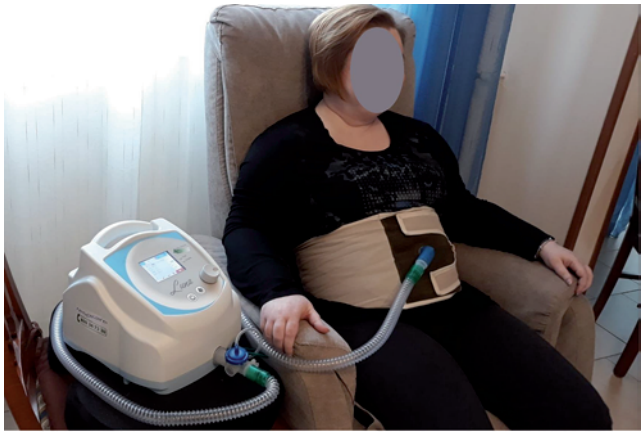

Figure 1. IAPV - Luna device (Dima, Bologna, Italy).

Follow-up and outcomes

At baseline (after 60' without ventilation in spontaneous breathing) and after one hour of IAPV, we recorded ABG and respiratory param-

Table 1. Patient's data at admission (T0) after 60' without ventilation in spontaneous breathing.

Arterial blood gas analysis	
pH	7.52
$PaCO_2$ (mmHg)	49
PaO_2 (mmHg)	46
HCO_3 (mEq/L)	36.3
BE (mmol/L)	11.3
Spirometry	
FEV_1 (%) L.	0.51 (18%)
FVC (%) L.	0.74 (21%)
Night-time polysomnography in NVS	
SpO_2 average %	94.2
SpO_2 nadir %	93
T90 %	0%
ODI	4.4/h
AHI	4.8

$PaCO_2$, partial pressure of carbon dioxide; PaO_2 partial pressure of oxygen; FEV1, forced expiratory volume in the first second; FVC, forced vital capacity; NVS, non-invasive ventilation support-setting; T90, time spent with SpO2 <90%; ODI, oxygen desaturation index; AHI, apnoea-hypopnea index.

Table 2. At the beginning of the adaptation to intermittent abdominal pressure ventilation. Comparison of gas exchange and respiratory parameters at baseline after 60' without ventilation in spontaneous breathing and after 1 hour during IAPV.

	SB	T0 IAPV (1 hour)
pH	7.49	7.44
$PaCO_2$ (mmHg)	46	33
PaO_2 (mmHg)	52	84
HCO_3- (mEq/L)	34.2	26
BE (mmol/L)	10.3	8.2
RR (cpm)	32	15
IV (mL)	385	630
EV (mL)	300	667
PIF (lpm)	35	47
PEF (lpm)	28	53

SB, spontaneous breathing; IAPV, intermittent abdominal pressure ventilator; BE, base excess; RR, respiratory rate; IV, inspiratory volume; EV, expiratory volume; PIF, peak inspiratory flow; PEF, peak expiratory flow.

eters (Table 2). The patient was discharged with instructions to continue daytime ventilation at home with IAPV for at least 10 h/day. Six months later, IAPV was still effectively relieving her daytime dyspnea, reaching up to 6 h/day of autonomous breathing without dyspnea or tachypnea and her BMI was 28.2 kg/m^2. After 18 months, IAPV efficacy persisted and the woman continued to use IAPV twice a day, for a total of 5 hours/day and it was well tolerated.

Table 3 shows the respiratory gas exchange measured with a blood gas analysis at baseline, at 6 and 18 months, after 60' without ventilation in spontaneous breathing, with overall improvement. Finally, to monitor the patient's quality of life and psychological well-being during adaptation to IAPV, an assessment was conducted by a psychologist at baseline, 6 and 18 months. At the time of discharge, the patient shows significant improvement in anxiety, depression, and quality of life (Table 4).

Discussion

NVS is important life-support commonly used in patients with neuromuscular diseases [1-3] both in acute and chronic respiratory failure [5,6]; it has been available for many years and different methods have been proposed. Usually, NVS is used with a variety of interfaces and settings, guarantying the best comfort for the patient [1,2]. Also, NVS includes body ventilators, tank ventilators, or iron lungs, but they are rarely used in Europe. IAPV, such as PBAir® is a valid alternative to NVS.

There are few case reports and fewer papers about the use of abdominal ventilation in clinical practice that show that the Pneumobelt® has the same effectiveness and safety as conventional ventilation devices [4,7-10]. Yang *et al*. showed the Pneumobelt® in night-time use in three patients with chronic respiratory insufficiency secondary to syringomyelia, poliomyelitis, and Friedreich's ataxia, respectively. It was found to be safe and effective in maintaining the daily function if used at night [7]. Hill *et al*. reported that Pneumobelt® use is effective in a sitting or standing position at an angle of 30° to 75° and during sleep. Its use is limited only to patients who can remain sitting in an upright position [11]. Moreover, there is a fitting problem, and its use is not effective in obese patients. In our case, we demonstrated that the IAPV corset is lightweight, comfortable and, thanks to Velcro fasteners, easier to fit and its use is possible in an obese patient as well. IAPV can improve verbal communication, appearance, and lower side effects. Only two patients developed sacral decubitus using the IAPV 24 hours a day [3].

In their retrospective study, Bach evaluated 209 patients who used the IAPV: 31 with a time of ventilator-free greater than 2 h, preferring mouth NVS. 178 patients with a time of ventilator-free less than one hour failed the IAPV trial and 50% of these patients had severe scoliosis. Fifty-four patients used the IAPV for long-term, and 48 used the IAPV for daytime support for a mean of 12.9±11.5 years [3].

Banfi *et al*. reported IAPV in a patient with chronic ventilatory insufficiency post-ischemic cervical myelopathy, who was dependent on continuous non-invasive ventilator support (CNVS) during sleep. Diurnal use of IAPV improved ABG values and the patient's mood and quality of life. However, he did not tolerate the CNVS facial interfaces during the day [9]. Moreover, De Mattia *et al*. used IAPV in a tracheostomized amyotrophic lateral sclerosis (ALS) patient for speech, and IAPV permitted optimal speech and guaranteed an efficient ventilatory pattern, with adequate gas exchange, without dyspnea [10].

Therefore, we can affirm the efficacy and safety of IAPV, as per the introduction of IAPV in a neuromuscular patient with LGMD who did not tolerate daytime NVS with any type of interface. In our case, the woman tolerated the new daytime device and it guaranteed optimal comfort, without any facial interface, leaving her free from facial skin necrosis. Thanks to IAPV, gas exchange improved, reaching the best ABG values for the patient and dyspnea diminished. These made it possible to reduce diurnal ventilation hours to at least 5/6 hours during the day, in two cycles.

As a result of these improvements, the patient interrupted continuous NVS, maintaining it at night and decreasing the hours of daytime ventilation with the support of IAPV, which was found to be well tolerated. Bach et al. stressed that regular follow-up is important because IAPV can become less effective with time [3]. The last follow-up was at 18 months after starting IAPV. Gas exchanges were

Table 3. At the beginning of the adaptation (T0), at T1 and T2, after 60' without ventilation in spontaneous breathing.

	Baseline (T0) SB	After 6 months (T1) SB	After 18 months (T2) SB
pH	7.52	7.42	7.42
pCO_2 (mmHg)	49	42	38
pO_2 (mmHg)	46	85	93
HCO_3- (mEq/L)	36.3	28.5	24.6
BE (mmol/L)	11.3	2.4	0.1

Table 4. Psychological monitoring.

Job activity	Psychological tests Baseline	Bank employee Post (6 months)	Follow-up (18 months)
EuroQoL (EQ-5D)	33311	33312	33312
EQ-5D Index	-0.429	-0.581	-0.581
Visual Analogue Scale (VAS EQ-5D)	60	20	15
World Health Organization Quality Of Life (WHOQOL)	78	76	88
WHOQOL - Physical domain	14 —> 62.5	10.66—> 41.66	16—>75
WHOQOL - Psychological domain	12.8 —> 55	12.8—> 55	14.4—>65
WHOQOL - Social domain	18 —> 87.5	18—> 87.5	18—>87.5
WHOQOL - Environmental domain	18.66 —> 91.6	19.33—>95.83	20.66—>104.16

maintained at the improved levels (Table 3) and the patient's tolerance and satisfaction reinforced this result (Table 4).

Conclusions

This case report showed the effectiveness, tolerability, and safety of IAPV in a patient who did not tolerate the standard daytime NVS. Despite the evidence of its safety and effectiveness, there are still no current guidelines for IAPV in neuromuscular disease and it remains an unusual alternative to NVS.

IAPV, in selected patients, should be considered an effective and safe alternative to guarantee a correct ventilatory pattern when NVS is not well tolerated or its interfaces generate discomfort in the patients, thereby leading to a better quality of life.

Further studies are needed to evaluate IAPV tolerability, safety, and efficacy in neuromuscular disease.

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