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Combined high flow nasal cannula and negative pressure ventilation as a novel respiratory approach in a patient with acute respiratory failure and limb-girdle muscular dystrophy

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We describe the case of a 56-year-old-man with limb-girdle muscular dystrophy affected by acute hypercapnic failure secondary to pneumonia treated with high flow nasal cannula, intermittent abdominal ventilation, and negative pressure ventilation. The patient did not tolerate noninvasive positive pressure ventilation and refused invasive ventilation and tracheostomy. We successfully experienced a novel approach combining high flow nasal cannula with cycles of intermittent abdominal pressure ventilation and negative pressure ventilation.

Key words: high flow nasal cannula, intermittent abdominal pressure ventilation, negative pressure ventilation, limb-girdle muscular dystrophy

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Abbreviations

HFNC: High flow nasal cannula
IAPV: intermittent abdominal pressure ventilation
NPV: negative pressure ventilation
NVS: noninvasive ventilatory support

Introduction

NVS (Noninvasive Ventilatory Support) is usually the therapy of choice for chronic respiratory failure in patients with muscular dystrophies, but some patients are intolerant of its use. In such patients, the high flow nasal cannula (HFNC) supportive therapy has emerged as a safe, useful therapy in acute and chronic respiratory failure, improving oxygenation and comfort. There are no data on the use of HFNC in patients with neuromuscular diseases.

Respiratory involvement is an almost constant feature of several muscular dystrophies, in particular of Duchenne muscular dystrophy (DMD) but also of some types of Limb- Girdle-Muscular-Dystrophy (LGMDs) and congenital types. Respiratory muscle weakness develops insidiously during the disease and patients need support that HFNC alone (Fisher & Paykel Healthcare AIRVO 2) cannot guarantee.

We now report a case of hypercapnic respiratory failure with intolerance to NVS where the alternate association of HFNC with cycles of intermittent abdominal pressure ventilation (IAPV with LunaBelt Dima Italia and negative pressure ventilation (NPV), PegasoVent DimaItalia with nylon poncho surrounding semi-cylindrical tent-like support) was successful and well-tolerated.

Case presentation

A 56-year-old-man affected by LGMD still in genetic staging, having acute respiratory failure, refused intermittent NVS because of intolerance, claustrophobia, and psychological reasons (fear of not being able to call family).

The patient was hospitalized with acute respiratory failure. At first, he was sedated and treated with NIV in pressure support ventilation with parameters: positive end-expiratory pressure 5 cm H_2O, pressure support 10 cm H_2O, respiratory rate 14/min, the fraction of inspired oxygen (FiO_2) 63%; Arterial Blood Gas (ABG) showed pH: 7,45, pCO_2: 43 mmHg, pO_2 93 mmHg, Lac: 1,1, HCO_3^- 30,1 Mmol/l.

Clinical condition and ABG remained stable for two days during NIV, but after the onset of a nasal pressure sore, the patient became shaken and no longer tolerant to NIV, with worsening of ABG parameters (FiO_2 60%, pH: 7.30, pCO_2: 80 mmHg, pO_2: 78 mmHg, HCO_3-: 35.3 mmol/L).

We tried to replace the old mask with one that avoided nasal lesions, but the patient categorically refused to wear any type of mask.

The patient refused intubation and tracheostomy, too. Therefore, we started the HFNC with the following parameters: flow: 60 L/min FiO2 60%, temperature 37° and IAPV (Pressure belt: 20 cm H_2O; T inspiratory: 1,3 sec; respiratory rate: 14 bpm; rise time: 0,6 sec).

After 1 h of this ventilatory approach, ABG parameters improved: pH: 7.43, pCO_2: 63 mmHg, pO2: 86 mmHg, HCO_3^-: 41 mmol/L. Alternation of cycles of HFNC - with a reduction of FiO_2 to 50% -and of IAPV lead to a stabilization of ABG parameters: FiO_2 50%, pH: 7.47, pCO_2: 56 mmHg, pO_2: 68 mmHg, HCO_3-: 37.4 mmol/L.

After 48h, the patient refused the IAPV treatment, so we alternated HFNC (Flow: 60 L/min, FiO2 50,% T 37°C) with NPV (PI: 40; PE: -01; F:14 1/E: 2.1:1) for 3/day of 3 hours per cycle getting a significant improvement of ABG: (FiO_2 50%, pH: 7.47, pCO_2: 51 mmHg, pO2: 76 mmHg, HCO_3^- std: 31.7 mmol/L).

In the subsequent weaning, only NPV guaranteed support. Table I shows an arterial blood gas. The patient tolerated the latter treatment well and agreed to continue NPV at home. He reported more comfort and a feeling of better control over his condition.

Discussion

Limb-girdle muscular dystrophies are progressive muscular diseases in which respiratory complications may be one of the main causes of death [1]. NVS should be the standard of care for respiratory support in patients with muscular dystrophies with survival benefit and upgraded quality of life. Many patients do not tolerate NVS and may have some episodes of acute respiratory failure during their disease with the risk of intubation or tracheotomy [2]. NVS intolerance is one of the major elements for high intubation rates [3]. Decubitus lesions, claustrophobia, or fear of not being able to call for help, can cause rejection or failure of NVS.

There are at least two reasons for not using dry oxygen via a nasal cannula: the FiO2 used is initially too high, and also the low-flow oxygen predisposes to epithelial lesions and dryness of the mucous membranes leading to scabs and crusts formation, some discomfort

Table I. Sequential ABG values.

Parameter	NIV (FiO_2 63%)	Ventimask (FiO2 60%)	1 hour: HFNC (FiO_2 60%-flow rate 60 L/min+IAPV	After 24 hour: (FiO_2 50%-flow rate 60 L/min+IAPV	1 hour: HFNC (FiO_2 50%-flow rate 60 L/min+NPV
pH	7.45	7.30	7.43	7.47	7.47
pO_2 (mmHg)	93	78	86	68	76
pCO_2 (mmHg)	43	80	63	56	51
SO_2 (%)	100	94.8	95.7	93.2	96
Lac (mmol/L)	1.1	1.3	0,6	1.1	1.3
HCO_3^- (mmol/L)	30.1	35.3	41	37.4	31.7

ABG: arterial blood gas analysis; PO_2: partial pressure of oxygen; PCO_2: partial pressure of carbon dioxide; SO_2: oxygen saturation; Lac: lactates, HCO_3^-: Bicarbonate

and irritation. Sometimes, it can also cause nosebleeds [4]. HFNC oxygen treatment is effective in the management of adults with acute hypoxemic respiratory failure, and to a minor extent, in patients with acute hypercapnic respiratory failure or weaning [5,6]. One of the major effects of HFNC in the nasopharynx is to wash CO_2, which reduces dead space and increases the ratio of alveolar ventilation to minute ventilation, decreasing resistive work of breathing. The dead-space wash-out, nasopharyngeal resistance reduction, positive pharyngeal pressure, alveolar recruitment, oxygen dilution reduction, decreased work for breathing, and patient comfort. In muscular diseases with reduced functionality of the central drive, there is no sign of the use of HFNC. Despite the benefit of oxygenation and the small level of pressure generated, when muscle breakdown is advanced, HFNC cannot be used alone as the oxygen. Using HFNC during IAPV, and the use of a combined system, may enhance the advantages of both techniques. IAPV comprises an elastic inflatable bladder incorporated into a corset surrounding the abdomen. Through a ventilator which inflates bladder, the abdominal content and diaphragm move upward, assisting the expiration phase. With bladder deflation instead, the inspiration occurs passively. Only scattered reports on the use of IAPV [7] are available and only one paper concerning its use in large populations of patients, in a regimen of noninvasive ventilator support [8]. IAPV facilitates diaphragmatic motion and may be useful in patients with bilateral diaphragmatic weakness or paralysis. Negative pressure ventilation was successfully and predominantly used for long-term mechanical ventilation until the mid-1980s. Later, the interest waned, partly because the noninvasive positive pressure ventilation has proven to be more effective in patients with altered pulmonary or chest wall mechanics, and in those with obstructive sleep comorbidities apnea-hypopnea. Technological evolution has developed small and portable devices, while NPV needs a poncho, an interface more voluminous than a mask. NPV is a ventilation model in which sub-atmospheric pressure during inspiration affects chest surface, which determines the expansion of the thorax and a pressure decrease in the pleural space. This creates a pressure gradient that allows air to move from the airways to the alveolus. When the pressure around the thorax becomes less negative, the expiration takes place passively, thanks to the return of the lung and the rib cage [9].

IAPV was used in ALS tracheotomized patients to facilitate speech [10], and NPV is currently used in Duchenne muscular dystrophy patients [11] and in post-polio syndrome [12]. As far as we know, this is the first case of combined application of IAPV or NPV, in patients with limb-girdle muscular dystrophies.

Ethical consideration

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Conflict of interest

The Authors declare no conflict of interest.

Author contributions

PI and AA conceptualized the study, performed a literature review and drafted the manuscript. PI and LM performed a literature review and drafted the manuscript. AA and LM performed a literature review and collected data. GF critically revised the article. All authors read and the final manuscript.

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