

MPV promote adherence to nocturnal NIV in a Duchenne patient

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We described a case of a patient 20 years old, affected by Duchenne dystrophy with obstructive sleep apnoea syndrome and severe nocturnal desaturation. He was not compliant to non-invasive ventilation (NIV) for claustrophobia and panic attacks. Mouthpiece ventilation was successfully used in this patient, who later accepted the nighttime NIV.

Key words: Mouthpiece ventilation, Duchenne dystrophy, NIV

Introduction

Non-invasive ventilation (NIV) is sometimes considered suboptimal in neuromuscular disease patients, due to excessive secretions in the airways, hypercapnia due to inadequate adherence, or lack of tolerance of the interface ¹. Interfaces that cover the nose and/or mouth and nose, are the most commonly used but they often cause skin lesions and some times even claustrophobia. Mouthpiece ventilation (MPV) was first described in 1953 to permit iron lung users without autonomous ability to breathe, to leave iron lungs during the day. The interface is provided by angled mouthpieces of 15 or 22 mm accessible to the mouth by means of a flexible support arm so that the patient can access them with the lips as needed. Patients can trigger the breath by creating a small negative pressure or by kissing the mouthpiece ². The main advantage over a nasal or oronasal mask is that mouthpiece produces less interference with language, no risk of skin breakage and claustrophobia. It is safer by allowing the use of glossopharyngeal respiratory ventilator failure or accidental disconnection from the ventilator.

Case report

In a 20 years old patient, affected by Duchenne dystrophy and hypokinetic dilated cardiomyopathy with moderate reduction of contractile function, and mild to moderate mitral and tricuspidal insufficiency, a sleep apnoea obstructive syndrome with severe nocturnal desaturation was diagnosed in the last year and use of CPAP was recommended to him. Though numerous interfaces were attempted to adapt him to the CPAP, the patient refused it for claustrophobia and the onset of panic attacks. One year later, the patient began to experience headache and morning tiredness; soon after he was admitted to the emergency room due to acute neurological symptoms, such as tremor, allucinations and dizziness. Clinical examination and blood test were normal. Blood gas analysis showed hypercapnia (52 mmHg) and mild hypoxemia (61 mmHg), requiring non-invasive mechanical ventilation. The patient was in a state of agitation, frightened and

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Conflict of interest

The Authors declare no conflict of interest

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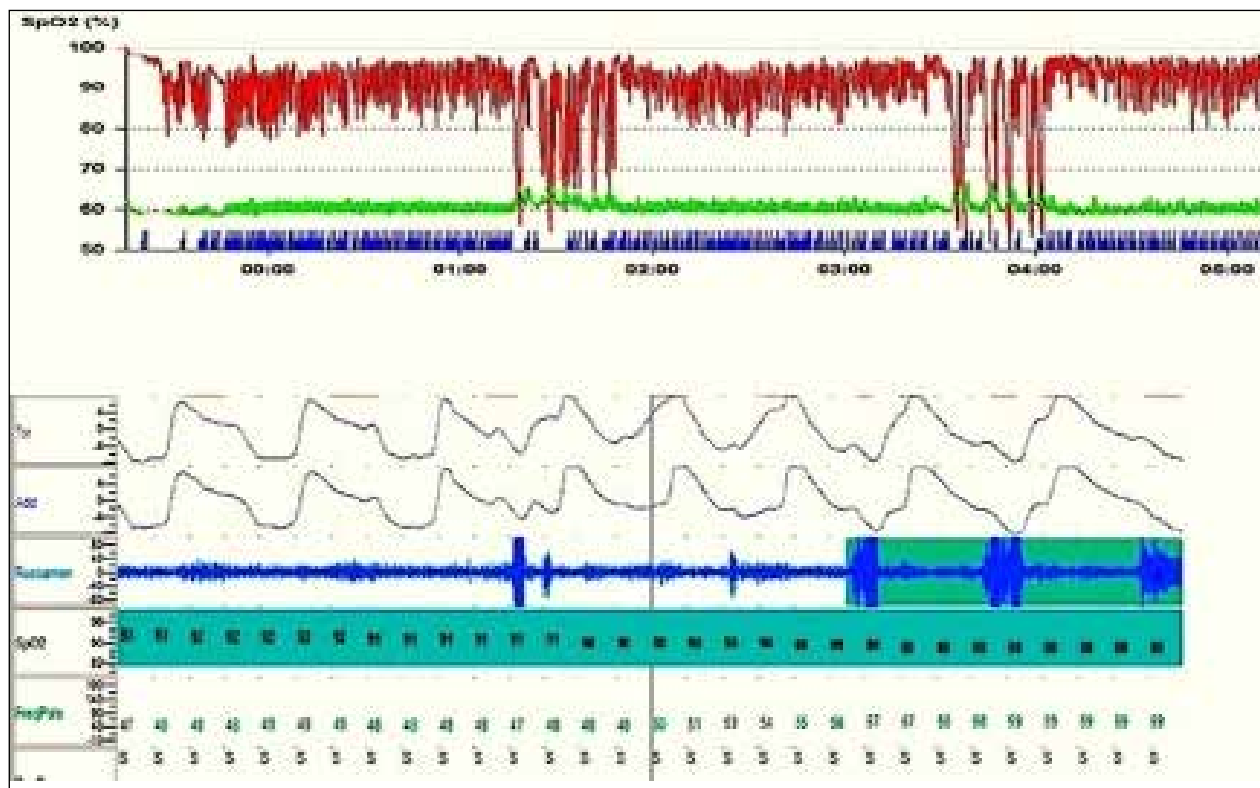


Figure 1. Overnight Polysomnography showing desaturation.

claustrophobic, but fully aware and refusing non-invasive mechanical ventilation with traditional interfaces. At this time, we proposed him MPV. The patient accepted to try the mouthpiece.

The inspiratory pressure was set between 12 and 14 mmHg, which obtained an optimal tidal volume between 8-10 mL/kg; No back-up rate was needed for daytime use, so no air blew into the patient's face; the inspiration time was set at 1.2 sec, EPAP 0, the rise time at 2/6. After only 1 hour, we observed an improvement in gas analysis values with normalization of CO₂ (46 mmHg) and oxygen (71 mmHg) values. The overnight polysomnography confirmed the presence of the severe obstructive sleep apnoea syndrome and prolonged hypoventilation (Fig. 1).

During the day, the patient selected MPV position with a rigid arm fixed on the bed or wheelchair. After only 2 days of MPV during daytime hours, the patient felt better and safer than before so that he accepted night ventilation with a nasal mask (Fig. 2).

He was adapted with mode ST-AVAPS, and parameters were the following: IPAP max 14 min 10, EPAP 6, Vt 650 mL; trigger flow 2.0 litres, 25% cycle, rise time 3, respiratory rate bk 10/min, Insp. Time 1.2 sec. Nocturnal pulse oximetry was performed, which confirmed the



Figure 2- The patient during MPV.

resolution of the desaturation events (Fig. 3). Blood gas analysis on awakening was normal.

Discussion

In this case report, the main goal of nocturnal NIV was achieved with the use of MPV. The use of MPV has been

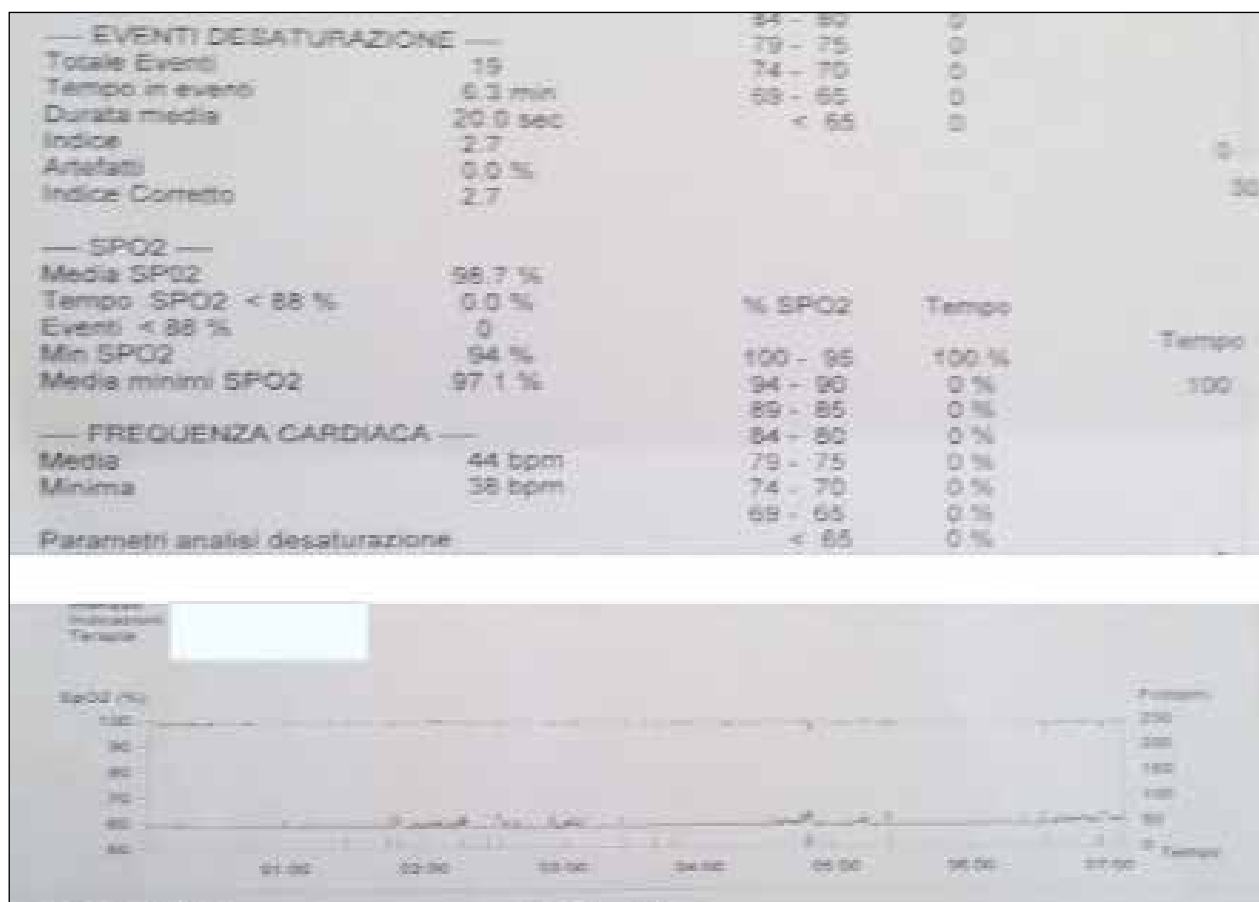


Figure 3. Pulse oximeter showing the correction of desaturation.

known for many years, but its diffusion has always been minimal until 2013 when models dedicated to commercially available portable ventilators were introduced. Despite this, still today, there is little knowledge of how to use it.

Bach et al.³ reported the sequential use of mouthpiece during the day and a nasal mask during the night. They also suggested the possible use of a standard mouthpiece with lip-seal retention or custom-moulded orthodontic bites for overnight use.

Positive expiratory pressure (EPAP) cannot be maintained for patients who use open NIV system, and is indeed rarely, if ever, necessary for these patients. Apnea alarms, when present, should be set at the highest threshold to avoid unnecessary activation and discomfort. The most common ventilator mode used is assisted volume- and pressure-controlled with no EPAP, low-pressure alarm set to apnea minimum and maximum duration⁴.

It is always necessary to carefully monitor the patient during the adaption phase since MPV requires true collaboration from the patient, and not all ventilators ensure rapid adaption to the patient's respiratory acts⁵. However, due to its specific features and disadvantages (air leaks,

etc.), MPV must be managed by expert hands and in a well-monitored way⁶. The use of MPV is likely limited to a few centres, for the longest time required to adapt and monitor the patient. The ventilator is easily activated by the mouth pressure of the patient. The mouthpiece is a preferable and comfortable alternative to NIV, but a more active participation is needed compared to the use of traditional masks.

It should always be considered for patients with chronic disease who need to start NIV; it is helpful to promote a positive approach to NIV. Patients reported a decrease in breathlessness and an increase in general and social productivity.

Our opinion is that it is necessary to know all the treatment possibilities to offer the best and personal therapy to each patient. It is also necessary to keep in mind that these patients often have psychological problems from seeing relatives or friends die of the same disease. Therefore, the time when NIV begins, that usually coincides with the progression of the disease, is a crucial time for the patients which often determines their quality of life and would require adequate psychological support⁷.

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