

# Non-invasive mechanical ventilation as an alternative respiratory support during gastrostomy tube placement, in a patient with Duchenne muscular dystrophy, 24/24 hours ventilation dependent

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**Patients with Duchenne muscular dystrophy may benefit from gastrostomy tube feeding due to progressive dysphagia and malnutrition. However, due to concomitant pathologies, they are often at high risk for anesthesiologic complications. We describe how the non-invasive mechanical ventilation has been an alternative successful respiratory support option during the gastrostomy tube placement in a patient with Duchenne muscular dystrophy, on continuous NIV treatment. This report confirms how the use of NIV can support alveolar ventilation, before, during and after mini-invasive procedures, and prevent respiratory complications.**

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## Introduction

Patients with Duchenne muscular dystrophy (DMD) may have complications such as dysphagia and progressive weight loss during the end-stages of the disease. As a result, aspiration pneumonia, malnutrition up to severe cachexia, may arise<sup>1</sup>. Often at this stage, patients are also dependent on non-invasive mechanical ventilation (NIV) due to severe muscle weakness. In these conditions percutaneous endoscopic gastrostomy (PEG) becomes necessary to improve the nutritional status of patients. Unfortunately, patients and their families accept PEG only late, when oral feeding becomes impossible due to dependence on continuous NIV.

In patients with neuromuscular pathologies, and especially in those suffering from severe DMD undergoing invasive procedures, it is desirable to minimize sedation to preserve the respiratory function, often already compromised. It should be remembered that anesthetic procedures in such patients become very complex due to difficulty in emergency intubation, and if intubated, the high risk of weaning failure due to invasive venti-

lation<sup>2</sup>. Tracheostomy may be required and/or serious complications such as ventilator-associated pneumonia may occur.

To our knowledge, there are only few reports in literature on the use of NIV in patients with DMD during PEG<sup>3-5</sup>.

We describe a 28-year-old patient with Duchenne muscular dystrophy in 24/24h non-invasive mechanical ventilation with a nasal mask, undergoing PEG with the support of NIV.

## Case report

Patient 28-year-old, affected by Duchenne muscular dystrophy, chronic respiratory failure and dilated cardiomyopathy (ejection fraction 40%), in treatment with ACE-inhibitors, warfarin, deflazacort, anti-oxidants and micro-nutrients. Despite supplementary nutritional support, the patient continued to lose weight due to the difficulty in eating orally. He was therefore offered to place a PEG, which he categorically refused. During the last 14 months his weight has further reduced from 48 to 35 kg, so much so that the patient himself was finally convinced to ask for PEG. At the time of the admission at our Unit, the serum level of prealbumin was 9.5 mg/dl (normal, 16.7-29.6 mg/dl), indicating severe malnutrition, while forced vital capacity (FVC) was 180 ml (7% of predicted, based on arm-span height estimate). Prior to the procedure, warfarin was replaced with sub-cutaneous low molecular weight.

### PEG placement

The anesthetic evaluation assessed the patient as a high-risk subject (ASA IV), with a high probability of tracheal intubation. The patient was sedated with midazolam, 2 mg before and 4 mg during the procedure, with 5' infusion rate.

The overall duration of the procedure was approximately 15 minutes. The patient was ventilated through the controlled assisted ventilation (CAV) mode with 450 ml tidal volume, and 14 back up respiratory rate. During the procedure the tidal volume was increased to 650 ml and the back up rate to 16, to compensate for the losses due to the placement of a mouthpiece for a few minutes to allow the passage of the gastroscope (Fig. 1), a greater alveolar recruitment, and the achievement of 95-96% stable peripheral saturation.

The following clinical parameters were monitored throughout the procedure: heart rate, blood pressure, electrocardiogram, and SpO<sub>2</sub>. Chest lift was also frequently monitored.

Respiratory support was supplemented with oxygen, 1-2 liters per minute, to avoid sudden desaturations. Tidal



**Figure 1.** Patient during percutaneous gastrostomy tube placement.

volume was titrated to achieve a good chest lift and air exchange, compensating for circuit losses and partial upper airway obstruction.

The procedure was successfully completed. At the end of the procedure, the ventilatory parameters were progressively reduced until the initial parameters were restored; the patient continued usual NIV during the post-operative phase and transport to the room. The gastrostomy tube was used for feeding, after 24 hr.

## Discussion

In end-stage Duchenne muscular dystrophy, a balanced nutritional approach is essential for patient care. However, in the phase of transition to wheelchair, over-nutrition often occurs caused by reduced caloric needs associated with reduced physical activity and energy expenditure at rest<sup>3</sup>. The excess caloric intake may also be due to the use of medication (steroids) with a consequent increase in appetite and/or compassion on the part of the parents, who tend to gratify the patient with food<sup>6</sup>. This results in an increased risk of obesity, dyslipidemia, hypertension, and often obstructive sleep apnea. As the disease progresses, loss of muscle strength and dysphagia are the main causes of malnutrition. Gastrointestinal problems (constipation, delayed gastric emptying, gastric reflux) may also arise, resulting in reduced food intake<sup>6</sup>. Prolonged meals and dependent eating are all consequences of muscle weakness and gastrointestinal dysfunction. The presence of respiratory insufficiency, constant in the advanced stages of the disease, further increases energy expenditure.

For these reasons, as respiratory failure progresses and weight loss begins, it is very important to evaluate the usefulness of nutritional support and PEG placement early. Over time, the patient's clinical condition may worsen, respiratory failure may evolve, and the patient may become dependent on NIV, making the endoscopic procedure more complex and life-threatening.

The case here reported is the first case in the litera-

ture describing support with NIV in a patient with DMD, chronic respiratory failure (FVC < 10%), dependent on NIV 24/24 per day.

A case of a patient with DMD and 17% FVC, in NIV, was previously described<sup>3</sup>. The procedure in that case was extremely complex and several attempts were made to settle the patient. The NIV had failed and an alternative method of ventilation was therefore used, manually ventilating the patient through an anesthesia bag connected via a short tube to the patient's NIV mask, with 100% oxygen blended into the circuit. In this way, the anesthesiologist, adjusted the volume of gas delivered with the anesthesia bag to maintain a good chest lift and air exchange.

From a practical standpoint, it is essential to recognize that there is no low-risk method of assisted ventilation for subjects with severe muscle weakness who require sedation for medical procedures.

It is therefore imperative that doctors of patients with severe DMD be aware of this. Furthermore, if a procedure requires sedation, the potential benefits, such as possible prolongation of survival and improved quality of life, and possible risks, such as emergency intubation, potential extubation failure, and the risk of tracheostomy should be carefully and equally evaluated<sup>7</sup>.

The benefits of NIV in patients with DMD are recognized in terms of prevention of hospitalization, recovery of alveolar hypoventilation, treatment of acute or chronic secondary respiratory failure, weaning from oro-tracheal intubation, and last but not least increased survival.

The use of non-invasive mechanical ventilation in patients with pre-and post-operative support needs, anecdotally already reported<sup>3-5</sup> in literature, requires further studies to validate its use in extremely fragile patients, at high anesthetic risk.

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None.

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#### *Conflict of interest*

The Authors have no conflicts of interest to disclose.

#### *Author contributions*

Conceptualization and preparation of the manuscript: AA, GF; data collection: AA, RC, EDC, writing and editing AA GF, revision of the manuscript RC, EDC, supervision AA, GF.

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