

Non-invasive ventilation in neuromuscular disease with acute respiratory failure: A narrative review

Mia Elhidsi¹, Dyah Ayu Kusumoputri Buwono², Eka Musridharta³, Dicky Soehardiman⁴, Prasenhadi⁴

¹Department of Pulmonology and Respiratory Medicine, Faculty of Medicine, Universitas Indonesia, Persahabatan National Respiratory Referral Hospital, Jakarta, Indonesia

²Faculty of Medicine, Universitas Indonesia, Persahabatan National Respiratory Referral Hospital, Jakarta, Indonesia

³National Brain Center Hospital, Jakarta Indonesia

⁴Department of Pulmonology and Respiratory Medicine, Faculty of Medicine, Universitas Indonesia, Persahabatan National Respiratory Referral Hospital, Jakarta, Indonesia

ABSTRACT

Acute respiratory failure (ARF) is a life-threatening condition that often results from the acute onset of neuromuscular disease (NMD) and often coexists with other cardiorespiratory conditions. For more than two decades, existing studies have shown that non-invasive ventilation (NIV) is the main ventilatory support and provides a good clinical outcome in ARF with various conditions, but its use in NMD patients with ARF is still limited. In patients with ARF, NIV can be initiated in fully awake patients with hemodynamically stable, without upper airway obstruction, and airway secretions can be overcome. Furthermore, expiratory positive airway pressure (EPAP) and inspiratory positive airway pressure (IPAP) settings with backup rates are recommended. Additionally, some studies have reported that the application of NIV in NMD with ARF can also be beneficial as a weaning strategy. Hence, further studies need to be conducted to generate evidence regarding the role of NIV in NMD patients with ARF.

Keywords: Non-invasive Ventilation, Neuromuscular Disease, Acute Respiratory Failure

Abbreviation List:

AECOPD	Exacerbation of chronic obstructive pulmonary disease	NIV	Non – invasive ventilation
APCV	Assisted pressure control ventilation	NMD	Neuromuscular disease
BiPAP	Bilevel positive airway pressure	PaCO ₂	Partial pressure of carbon dioxide
BMI	Body mass index	PCO ₂	Partial pressure of carbon dioxide
CM	Cricothyroid “mini-tracheostomy”	PIMAX	Maximal inspiratory pressure
CO ₂	Carbon dioxide	Ppl	Pleural pressure
CO ₂ PD	Chronic obstructive pulmonary disease	PSV	Pressure support ventilation
CPAP	Continuous positive airway pressure	RCTs	Randomized controlled trials
DMD	Duchenne muscular dystrophy	SBC	Single breath count
EPAP	Expiratory positive airway pressure	SIMV	Synchronized intermittent mandatory ventilation
GBS	Guillain – Barré syndrome	Ti	Inspiratory time
ICU	Intensive care unit	VAPS	Volume – assured pressure support
IMV	Invasive mechanical ventilation	VC	Vital capacity
IPAP	Inspiratory positive airway pressure	WOB	Work of breathing
MEP	Maximal expiratory pressure		
MIP	Maximal inspiratory pressure		

INTRODUCTION

Acute respiratory failure (ARF) is a condition characterized by an abrupt loss in lung function, re-

sulting in inadequate gas exchange, arterial oxygen tension < 60 mmHg, normal or low arterial CO₂ tension, or increased arterial CO₂ tension >50 mmHg

Corresponding author:

Mia Elhidsi

E-mail: miapulmo.ui@gmail.com

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[1–3]. Acute respiratory failure is a life-threatening condition that often results from the acute onset of neuromuscular disease (NMD), which often coexists with other cardiorespiratory conditions. Variants of NMD have different risks of ARF. Studies have shown that Guillain–Barré syndrome (GBS) and myasthenia gravis are the two groups of NMD diseases that have the highest incidence of acute respiratory failure in developed countries [4]. Several factors, such as early onset of diagnosis, a high body mass index (BMI), pulmonary embolism, and pneumonia due to bacterial, viral, or aspiration pneumonia can increase the risk of ARF in NMD [5,6].

The management of ARF in NMD is comprehensive, including oxygen therapy, cough assistance, physiotherapy, antibiotics if needed, and intermittent positive pressure ventilation. NIV is ventilatory support that provides positive pressure into the lungs without invasive endotracheal tube intubation so that patients can be spared the complications associated with endotracheal intubation and conventional invasive mechanical ventilation (IMV) [7–9]. In more than two decades, existing studies have shown that NIV is used as the main ventilatory support in NMD and provides a good clinical outcome in ARF with various conditions, such as cardiogenic pulmonary edema, obstructive pulmonary disease (COPD) exacerbation, severe hypoxemia in immunosuppression conditions, and post-extubating from IMV [10–13].

However, to date, the use of NIV in ARF is still very limited. Furthermore, the indications for the use of NIV must be precise, and the relative contraindications also need to be considered. Therefore, we aimed to perform a narrative review to discuss the pathophysiology of ARF and the timing and applications of NIV in ARF with NMD.

PATHOPHYSIOLOGY OF ACUTE RESPIRATORY FAILURE IN NMD

In NMD patients, muscle weakness affects three muscle groups involved in the respiratory process: inspiratory muscles performing ventilation and voluntary inspiration, expiratory muscles, which contribute to forced expiration and forced expiration flow, and bulbar muscles [6,14]. The impairment of the diaphragm, intercostal muscles, and accessory muscles taken together can cause inadequate ventilation until micro-atelectasis occurs, contributing to subsequent ventilation/perfusion mismatch and hypoxemia [15]. Additionally, compensatory tachypnoea and an increase in work of breathing (WOB), which is often inadequate, may further exacerbate atelectasis and increase muscle fatigue. In this condition, the patient develops very high pleural pressure (Ppl) to generate airflow. At the same time, it causes a decrease in the respiratory system's ability to form a maximal inspiratory pressure (PI_{MAX}), which can increase the Ppl/PI_{MAX} ratio [4,6,14,15]. As

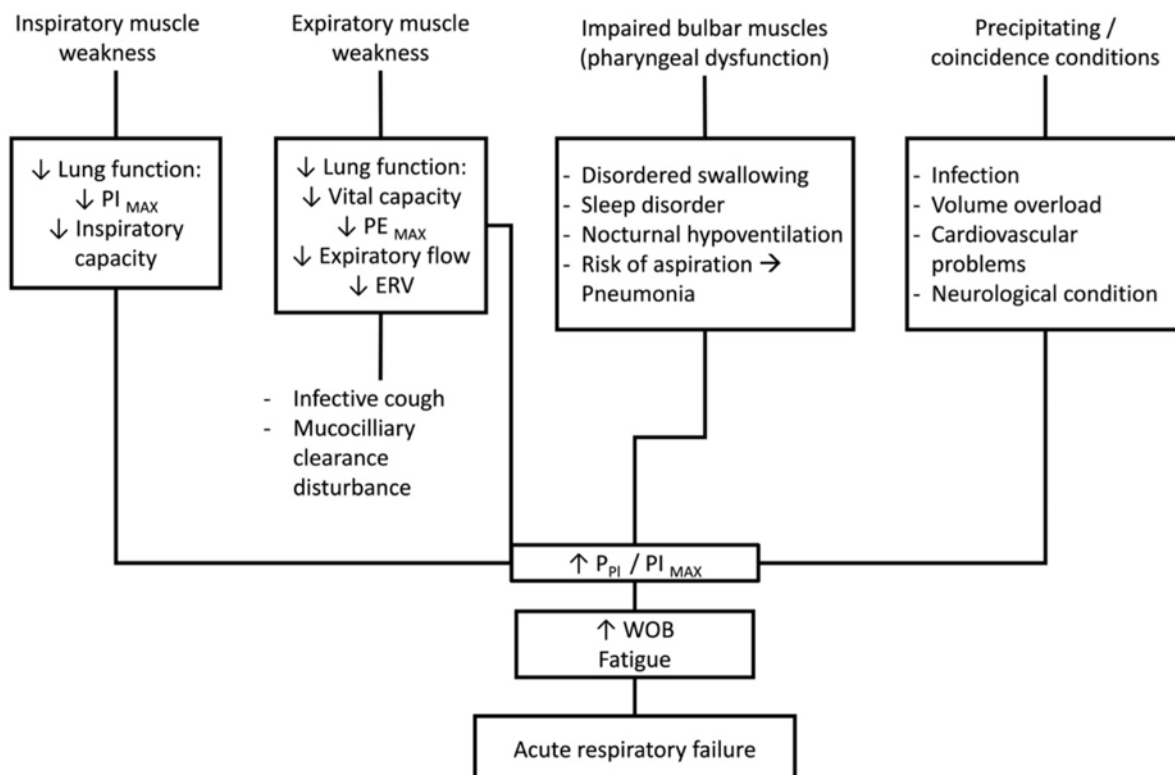


FIGURE 1. Pathophysiology of ARF in NMD patients

PI_{max} : maximum inspiratory pressure; PE_{max} : maximum expiratory pressure; Ppl: pleural pressure; ERV: Expiratory residual volume; WOB: work of breathing

fatigue increases, this condition triggers progressive hypercapnia, respiratory acidosis, and CO₂ retention in already weakened respiratory muscles. In the end, these circumstances tend to decrease the central respiratory drive (Figure 1) [5,15].

Weakness that occurs in the expiratory muscle group has the potential to interfere with the forced expiration process and airflow during the expiration process. Furthermore, this will lead to a failure of the airway clearance mechanism, encumbrance of airway secretion, and ineffective coughing, which consequently makes the patient more susceptible to respiratory tract infections, followed by complications of congested airway secretions [4,14]. On the other hand, impaired expiratory and inspiratory muscle contractility may be further aggravated by several other systemic involvements associated with NMD, such as decreased chest wall compliance due to the progression of scoliosis caused by atony of the paravertebral muscles. Furthermore, reduced chest wall compliance caused by progressive stiffness of the tendons and ligaments of the thoracic muscles may be a consequence of impaired muscle insertion caused by thoracic deformity [5,15]. Weakness of the bulbar and upper airway muscles may also increase the potential for mechanical obstruction of the upper airway and worsen the WOB. In addition, oropharyngeal and laryngeal muscle collapse may impede swallowing, speaking, regulating the cough mechanism, and intrathoracic pressure. In the supine position, it may increase the likelihood of swallowing and speech disorders, aspiration of airway secretions, and decreased ability of airway clearance, and increase the potential for recurrent respiratory infections [1,4,6,14].

Several circumstances, such as infection, volume overload, and cardiovascular and neurologic conditions, can trigger acute respiratory failure in NMD, although these precipitating factors alone can be a cause of acute respiratory failure [16]. Figure 1 describes the pathophysiology of acute respiratory failure in NMD.

NIV TIMING FOR ARF IN PATIENTS WITH NMD

The aim of ventilation administration in NMD patients with ARF is to improve gas exchange by unloading the work of respiratory muscles and encouraging cough augmentation to improve airway clearance and increase lung function parameters to prevent pulmonary physiological deterioration due to atelectasis and chest wall abnormalities. Even though the mechanisms by which NIV produces favorable results are not fully understood, certain studies have proposed some mechanisms. Possible mechanisms of how NIV could enhance oxygenation in NMD patients with acute respiratory failure

are by reducing respiratory muscles' work, resetting the respiratory center to a lower level of PCO₂, increasing pulmonary mechanical abilities, reducing bicarbonate retention in kidney, resetting body chemoreceptors, improving sleep physiology, and enhancing the central respiratory response to the body's CO₂ level [5,17].

In patients with ARF, NIV can be initiated in fully awake patients, and patent airways without upper airway obstruction and airway secretions can be overcome. Furthermore, NIV should be avoided in patients who are hemodynamically unstable [18]. In home NIV patients, adjustments are needed by increasing the NIV settings [16]. In addition, oxygen supplementation should also be given via the NIV to achieve a target oxygen saturation level 94-98% [19]. Oxygen supplementation without ventilation support is not recommended because it can reduce lung function and exacerbate hypercapnia and ventilation-perfusion mismatches. Monitoring cardiorespiratory, neuromuscular, and other precipitating factors should be carried out simultaneously. Moreover, the patient should be admitted to the ICU and moved to the intermediate room after the condition has improved [20].

The monitoring of patients' ARF on NIV should include clinical assessment of vital signs, work of breathing, level of carbon dioxide (CO₂) titter and neuromuscular progression within the first 2 hours after starting NIV. If oxygen level saturation fails to improve in addition to WOB, and there is no improvement in CO₂, endotracheal and invasive mechanical intubation should be considered [18]. Some studies have stated that it is preferable to perform endotracheal intubation as an elective procedure rather than emergency intubation. The decision is made based on clinical findings and objective measures. Pulmonary function tests consisting of indicators in the form of vital capacity (VC), maximal inspiratory pressure (MIP), and maximal expiratory pressure (MEP) can help the clinician predict a trend in patient respiratory function. In addition, single breath count (SBC) can also be used as a useful surrogate for VC in limited settings. In general, SBC < 20, VC < 20 mL/kg, MIP < 30 cm H₂O and MEP < 40 cm H₂O are some thresholds to use as a guide for elective intubation [21].

The use of NIV can also be beneficial in post-extubating NMD patients if the patient looks clinically weak, considering that the risk of re-intubation in NMD patients with ARF is quite high. Furthermore, NIV can be used continuously or at night (nocturnal NIV) to prevent airway and alveolar collapse [22]. Previous research has reported a good outcome in myasthenia gravis in GBS patients who have improved after extubating, especially when the patient is no longer experiencing dysautonomia and neuropathy [23,24].

NIV SETTING FOR ARF IN PATIENTS WITH NMD

The successful application of NIV depends on cooperation between the patient and the provider. In addition, some indicators potentially affect NIV success. The selection of the NIV mode and settings is based on the patient-specific diagnosis. The application of the two main modes of NIV, continuous positive airway pressure (CPAP) and bilevel positive airway pressure (BiPAP), to NMD requires special attention. Moreover, continuous positive airway pressure is not preferable in NMD patients due to respiratory muscle weakness and the increased work of breathing. In addition, NMD patients need extra force to resist the pressure during expiration, since CPAP only has continuous positive pressure and limitation in reducing respiratory muscle workload [25]. Furthermore, Bi-PAP is probably the most common mode in NIV and requires support from EPAP and IPAP to reduce the work of breathing during the expiratory phase [26]. However, the application of BiPAP in spontaneous modes without a backup respiratory rate is not recommended, since NMD patients often fail to trigger the device [25]. Currently, the use of advanced modes of NIV, such as volume-assured pressure, has been developed. Some studies recommend the application of volume-assured pressure support (VAPS), which can support the integration of both volume- and pressure-controlled non-invasive ventilation to ensure tidal volume in NMD patients [27–29].

Many experts recommend some considerations in NIV settings to enhance the likelihood of success in NIV application. It is suggested to set a target tidal volume of 6–8 ml/kg (ideal body weight). Moreover, EPAP should start at 4–5 cm H₂O, IPAP should start at a minimum of 5 cm H₂O, and the IPAP maximum level should be set high (10–20 cm H₂O above IPAP minimum). The larger the IPAP and EPAP, the greater the tidal volume that will be delivered. A backup respiratory rate is usually at 10–14 breaths per minute, since muscle workload is usually not powerful enough to trigger the ventilator reliably, especially during the sleep phase [16].

In addition, setting a high trigger sensitivity can also facilitate the patient's transition from the exhalation to inhalation phase, about 1–2 L/min. In a patient with a neuromuscular disorder, it is also recommended to set the inspiratory time (Ti) longer, at about 33 to 50 percent of the total respiratory cycle time, or 1.0–1.2 seconds, to ensure an adequate delivery of tidal volume per breath and a full inspiratory cycle [16,30]. Furthermore, it is also recommended to increase or extend the duration of NIV support in patients who are already using home NIV due to chronic respiratory failure, since there is no indication for intubation. In this condition, it is suggested to increase the IPAP level to provide a given tidal volume in 24 hours of NIV administration [16].

Proper fitting of the interface is another key indicator of successful NIV. Each interface has advantages and disadvantages in the utilization of NIV. To this point, no randomized studies have been performed to identify the most suitable interfaces in acute respiratory failure cases. Some main indicators regarding the choice of an interface are the optimization of more effective ventilation, the risk of air leakage, patient comfort, the size of the dead space, and the risk of upper airway obstruction. With NIV, upper airway obstruction may be caused by nasal obstruction, pharyngeal collapse, and/or glottic closure [31]. In the acute setting, well-fitted facial masks and oronasal masks are more commonly used and recommended, whereas nasal masks can also be used with a chin strap to prevent leaks [32–34].

CLINICAL STUDIES ON NIV IN NMD WITH ARF

Recently, some evidence has suggested NIV as ventilation support in cases of acute exacerbation of chronic obstructive pulmonary disease (AECOPD), cardiogenic pulmonary edema, de novo hypoxaemic respiratory failure, immunocompromised patients, chest trauma, palliation, post-operative care, weaning, and post-extubating patients [35,36]. However, these findings cannot be used as indicators to predict the success or failure of NIV prescription in neuromuscular disease patients with acute respiratory failure who also have some special features, consisting of facial weakness, airway secretion disorders, and bulbar muscle weakness, which may substantially extrapolate an NIV failure. Furthermore, Senevirame et al. showed that NIV effectiveness may be suddenly reduced by the presence of bulbar dysfunction, which may impede bronchial secretion clearance and cause excessive airway secretion [37]. Sudden worsening in respiratory function and vital signs can also occur in the use of NIV, thereby increasing the likelihood of emergency intubation [38]. In addition, a systematic review of randomized controlled trials (RCTs) or quasi-RCTs to compare the effectiveness of NIV and invasive mechanical ventilation for acute respiratory failure in neuromuscular disease and chest wall disorders conducted by Luo et al. did not identify any study eligible to be included in meta-analysis review [39].

Some observational studies have stated that NIV support may provide benefits for NMD with acute respiratory failure patients. Furthermore, a small retrospective study by Al Enezi et al. found an improvement in some respiratory parameters, including an increase in oxygen pressure level and pH level, and a decrease in CO₂ pressure, after the use of BiPAP [40]. Moreover, a case-control study compared 14 non-consecutive patients suffering from

ARF of neuromuscular origin who were administered NIV with 14 matched historical control patients receiving invasive mechanical ventilation, showing that the duration of ICU stay in the NIV group was shorter than in the IMV group. In addition, some of the subjects from the NIV group in this study also underwent a cricothyroid “mini-tracheostomy” (CM) [41]. In their prospective study of 17 patients with 24 acute respiratory failure episodes, Servera et al. found that 79.2% of the acute episodes could be prevented from intubation by using the NIV and cough assistance. This study found that bulbar impairment was an independent predictor for NIV failure [42].

A case series was reported by Kim et al. on NMD patients who had previously failed to wean according to the conventional weaning procedure due to muscle weakness and gradual hypercapnia. Additionally, the patients had to be reintubated, and then weaning was conducted using NIV. The study included a total of 18 patients, mostly Duchenne muscular dystrophy (DMD) patients, who were successfully extubated for at least five days. The patient was stable and had been discharged from the hospital alive [43]. Another study included an NMD patient who experienced acute respiratory failure and had NIV as a first-line ventilatory support. In this study, 42 of 51 patients used the NIV-assisted pressure-control ventilation (APCV) mode, with a full-face mask accompanied by a nasal mask. In this study, the success rate was quite high, with 36 pa-

tients (85.7%) being prevented from endotracheal intubation [44].

At the moment, we found a limited study with a small number of patients that provided clinical evidence regarding the application of NIV in NMD patients with acute respiratory failure. Consequently recommendations and conclusions are difficult to form. However, currently, the guidelines state that the NIV may be used in patients with ARF due to chest wall deformity or neuromuscular diseases ($\text{PaCO}_2 > 45$). Nevertheless, the use of NIV in weaning from IMV in NMD patients, as well as critical illness myoneuropathy, is challenging and needs to be monitored closely [36]. Moreover, there was low-quality evidence either pro or contra any of the weaning protocols proposed, involving using pressure support ventilation (PSV), synchronized intermittent mandatory ventilation (SIMV), CPAP, or a ‘T’ piece in weaning patients with NMD [45].

CONCLUSION

Acute respiratory failure in NMD patients often results from an exacerbation or coincidence with other precipitating conditions. Furthermore, NIV can be used as first-line ventilatory support in NMD patients with ARF who are fully awake, hemodynamically stable, without airway collapse, and without excessive airway secretions. Hence, further studies need to be conducted to generate evidence regarding the role of NIV in NMD patients with ARF.

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