

Case Report

Noninvasive Ventilation in Respiratory Type II Failure in a Severe Kyphoscoliosis Patient

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Abstract: Kyphoscoliosis, a severe chest wall deformity, can lead to chronic hypoventilation and type II respiratory failure due to compromised respiratory mechanics. Noninvasive ventilation (NIV) has become an effective therapeutic intervention in managing chronic respiratory failure secondary to neuromuscular disease (NMD) and thoracic cage abnormalities. We present the case of a patient with advanced kyphoscoliosis who developed acute respiratory failure (ARF) and was successfully managed with NIV. This case emphasizes the importance of individualized anticipatory care planning, appropriate NIV initiation, and optimal ventilator settings in patients with restrictive thoracic disorders. After NIV initiation the patients arterial blood gas has shown significant improvement. The initial plan of NIV included 2 sessions of two hours/day and during the whole night and at discharge the patient was recommended nocturnal NIV. The arterial blood gas analysis has shown a reduction of paco₂ from 62mmHg to 45 mmHg.

Keywords: Kyphoscoliosis; Type II Respiratory Failure; Noninvasive Ventilation; Chest Wall Disorders; Chronic Hypoventilation; Anticipatory Care Plan

1. Introduction

Patients with neuromuscular disease (NMD) and chest wall disorders such as kyphoscoliosis comprise a heterogeneous group, often showing significant variability in disease presentation and progression—even among those with the same diagnosis [1]. The same condition may present at different stages of its natural history, and long-term outcomes may be difficult to predict, particularly in patients presenting with acute respiratory failure (ARF) for the first time [2]. Although increased understanding of genotype–phenotype correlations in NMD has enhanced prognostic estimations, accurate forecasting of disease course remains elusive [2].

Compromised thoracic mechanics in kyphoscoliosis results in a decreased capacity of respiratory muscles, contributing to chronic alveolar hypoventilation and type II respiratory failure [3]. In recent years, noninvasive mechanical ventilation (NIMV) has gained popularity for managing chronic respiratory failure in these patients, leading to improved sleep quality, daytime activity, and overall quality of life [4,5].

There are few randomized controlled trials (RCTs) evaluating the use of NIV in ARF among NMD and chest wall disorder populations, making evidence-based guidance limited [6]. Nevertheless, early identification and individualized intervention remain essential. Anticipatory care planning is a cornerstone of this strategy—encompassing hospital versus home management, resuscitation preferences, and possible escalation to invasive ventilation [7]. These care plans should be regularly reviewed and updated based on disease trajectory, treatment response, comorbidities, and patient preferences [8].

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2. Results

A 47-year-old female with a known history of severe congenital kyphoscoliosis presented to the emergency department with increasing dyspnea, fatigue, and morning headaches. She had no prior history of mechanical ventilation or significant respiratory infections.

On examination, she exhibited tachypnea (RR 28), use of accessory muscles, and paradoxical abdominal motion. Arterial blood gas analysis showed a pH of 7.31, PaCO₂ of 62 mmHg, PaO₂ of 68 mmHg on room air, and bicarbonate of 30 mmol/L—consistent with type II respiratory failure. Chest radiograph revealed gross thoracic deformity with significantly reduced lung volumes, Cobb angle 72°. The spirometry evaluation has shown a severe restrictive disfunction: FVC 28 % predictive value.

Given the absence of sepsis or airway obstruction, she was initiated on NIV in the hospital setting using a pressure-targeted mode. The initiation of NIV was done according to the following criteria: a symptomatic patient with dyspnea, daytime hypercapnia and severe reduction of the FVC. Initial settings included IPAP 16 cm H₂O, EPAP 5 cm H₂O, backup rate 12 breaths per minute, and Ti of 1.0 s. The patient tolerated the interface (oronasal mask) well, and blood gases improved over 48 hours (PaCO₂ reduced to 49 mmHg). She was discharged home on nocturnal NIV with a follow-up plan involving home telemonitoring and outpatient physiotherapy. A nasal mask was recommended at home because of certain benefits for the patient—it allows the patient to cough and clear the oral secretions.

3. Discussion

This case reinforces the importance of early and individualized use of noninvasive ventilation (NIV) in restrictive thoracic disorders such as kyphoscoliosis. NIV should almost always be trialed in acutely unwell patients with NMD or chest wall deformity (CWD) who develop hypercapnia; importantly, clinicians should not wait for acidosis to occur before initiating NIV [9,10].

Face mask comfort is the first prerequisite for patient adherence to therapy. Some people report a sensation of claustrophobia when first introduced to mask ventilation. Several weeks of acclimation with gradually increasing duration of use may be necessary to overcome this experience. Skin surface discomfort, discoloration, inflammation, or frank skin ulceration are common, especially when higher inspiratory pressures are applied (). Our patient tolerated the initial NIV using an oronasal mask and at home a nasal mask was well tolerated.

This case emphasizes the importance of NIV for patients with severe kyphoscoliosis, and it is a example of the benefits of NIV in acute kyphoscoliosis-related respiratory failure.

Nasal masks are often recommended for initiation of nocturnal NIV in part because they free the mouth for cough, speech, and clearance of oral secretions or vomiting. Low-profile nasal pillow and cushion masks rarely cause skin pressure injury and are preferred by many patients over nasal masks that cover the nose. However, they may be easily displaced by lateral head movement, and they tend to leak at peak inspiratory pressures > 20 cm H₂O

The use of NIV in restrictive disorders is associated with multiple short- and long-term benefits, including improved gas exchange, hemodynamic function, sleep quality, and health-related quality of life [11]. The pathophysiological mechanisms by which NIV exerts its benefits may include respiratory muscle rest, modulation of central CO₂ sensitivity, and enhanced pulmonary mechanics [12].

In patients with early-onset scoliosis or other forms of CWD who retain expiratory and bulbar muscle function, NIV alone may suffice without the need for cough-assist techniques or secretion management interventions [13]. However, a combined approach—incorporating physiotherapy and mechanical insufflation-exsufflation—can be beneficial in patients with impaired cough efficiency [14].

Eligibility for NIV in these patients may be too conservatively defined. In response, updated German national guidelines now recommend NIV for patients with:

Nocturnal $\text{PaCO}_2 \geq 45$ mmHg,

Nocturnal transcutaneous CO_2 (PtcCO_2) ≥ 50 mmHg for over 30 minutes,

A nocturnal PtcCO_2 rise ≥ 10 mmHg above diurnal baseline in normocapnic patients,

A $>10\%$ decline in forced vital capacity (FVC) within 3 months for ALS or fast-progressing NMDs [15].

Sleep-related hypoventilation, especially during REM sleep, often precedes diurnal hypercapnia and may justify earlier initiation of ventilatory support [16]. Symptoms such as daytime hypersomnolence, morning headaches, dyspnea, and cognitive dysfunction should also trigger NIV consideration—even when resting PaCO_2 appears borderline [17].

Anticipatory care planning is critical to ensuring care decisions reflect the patient's values and medical condition. Such planning helps navigate decisions around hospitalization, escalation to invasive ventilation, ICU admission, and resuscitation preferences [7,8]. These principles apply broadly—from congenital muscular dystrophy to motor neuron disease and severe CWD—when care plans are individualized appropriately.

The setting of NIV initiation is another crucial factor. Home initiation of HNV, while less expensive and emotionally taxing, may be hindered by limited professional supervision and nighttime monitoring [18]. Hospital-based initiation offers increased safety, especially during the first NIV trials, but at the cost of greater financial burden and higher nosocomial infection risk [19]. Telemonitoring may offer a middle ground by enabling supervised home setup while maintaining safety.

Our patient's rapid clinical and arterial blood gas response following NIV initiation—reflected in a PaCO_2 decrease from 62 mmHg to 45 mmHg—parallels the short-term improvements reported in the literature. Specifically, Windisch W [4] reported that NIV improved blood gases and sleep architecture in patients with chest wall disorders, with an average PaCO_2 reduction of 10–15 mmHg over a similar timeframe. This consistency suggests that despite the heterogeneity of kyphoscoliosis presentations, appropriately initiated NIV can yield predictable physiological benefits.

In contrast with Bourke SC et al [5], which highlighted significant variability in tolerance and adherence to NIV due to mask discomfort and patient anxiety, our patient showed excellent tolerance to both oronasal and nasal masks. This may be attributable to the early anticipatory care planning, the careful selection of the interface, and stepwise acclimatization strategies used during hospitalization. Reference 5 also emphasized the challenge of maintaining long-term adherence, especially in home settings; our patient's adherence was supported by a structured discharge plan, telemonitoring, and physiotherapy—underscoring the importance of continuity of care.

The long-term benefits of NIV in restrictive disorders, including improved quality of life and survival, are well documented in Dreher M, et al [11]. Our case adds further real-world evidence to these claims by demonstrating not only gas exchange improvement but also symptomatic relief (e.g., reduced fatigue and morning headaches). Additionally, while Reference 11 emphasizes the role of NIV in chronic management, our case shows that early NIV use during an acute decompensation episode can also stabilize patients and avoid intubation.

Nasal interfaces, although not universally preferred, were well tolerated by our patients at home, aligning with data reported by Dreher M, et al (11) that suggest nasal masks are associated with fewer skin complications and improved comfort when inspiratory pressures are kept moderate.

Lastly, while Windisch W (4) reported a delayed initiation of NIV in some patients due to the absence of overt respiratory distress, our case supports the view that early intervention—guided by symptoms and hypercapnia even in the absence of acidosis—can prevent further deterioration.

4. Conclusions

Noninvasive ventilation is an effective and life-improving intervention in patients with kyphoscoliosis-induced type II respiratory failure. This case adds to existing literature by confirming the reproducibility of gas exchange improvements and symptom relief as documented in similar cases [4, 11], while highlighting how early initiation and tailored interface selection can mitigate some of the tolerance issues commonly described by Bourke SC et al [5].

Prompt recognition of chronic hypoventilation, timely initiation of NIV, and thoughtful interface and setting selection remain critical to success. Importantly, anticipatory care planning—emphasized throughout our management and consistent with current best practices—ensures preparedness for disease progression and escalated care if needed.

As the utility of home NIV expands, particularly with telemonitoring support, our case exemplifies how hospital-based initiation followed by outpatient continuity can combine the benefits of safety, personalization, and patient-centered care. These strategies, aligned with the evidence described by Windisch W [4], Bourke SC et al [5] and Dreher Met al [11] represent a growing consensus in managing restrictive thoracic disorders effectively and compassionately.

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