



Non-Invasive Ventilation in Severe Acute Respiratory Failure Complicating Madelung's Disease: A Case Series and Management Review

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Abstract

Madelung's disease (Launois-Bensaude syndrome) presents a critical challenge in managing acute respiratory failure (ARF) due to the high risk of difficult airway and failed intubation posed by symmetric cervical lipomatosis. This case series investigates the feasibility of non-invasive ventilation (NIV) as a primary strategy for ARF in these high-risk patients. We present two cases of severe ARF triggered by COVID-19 pneumonia in patients with Madelung's disease. Both patients were successfully managed with NIV using a total face mask, which avoided the need for hazardous endotracheal intubation. Key challenges included managing bilateral pneumothorax as a complication of NIV in one patient, and identifying and treating severe obstructive sleep apnea (OSA), a prevalent comorbidity as a crucial factor for successful weaning in the other. Our findings, integrated with a review of existing literature, demonstrate that a proactive NIV-first approach is a viable and potentially life-saving strategy in this population. This report provides a practical management framework, emphasizing careful interface selection, vigilance for complications, and systematic screening for underlying OSA to guide clinicians in optimizing outcomes for these complex patients.

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Introduction

Madelung's disease, also known as Launois-Bensaude syndrome, is a rare disorder characterized by the symmetrical, non-encapsulated accumulation of adipose tissue around the neck, shoulders, and upper trunk [1]. This unique anatomy creates a critical challenge in clinical practice: the management of acute respiratory failure (ARF). The fatty deposits can cause severe upper airway obstruction and distort oropharyngeal anatomy, classifying these patients as having a recognized "difficult airway" with a high risk of failed endotracheal intubation [2,3].

The convergence of this high-risk condition with the COVID-19 pandemic, which frequently leads to severe hypoxemic ARF, created a complex clinical scenario for which there are no established management guidelines. Non-invasive ventilation (NIV) presents a theoretically attractive strategy to avoid hazardous intubation, but practical evidence on its application in this population is scarce [4,5].

We present two cases that illustrate this dilemma. Both patients, with a known history of Madelung's disease, presented to the emergency department with acute hypoxemic respiratory failure. The primary symptom at presentation was severe dyspnea at rest.

Physical examination was notable for significant tachycardia, tachypnea, and, in one case, profound cervical dysmorphism with marked enlargement of the neck circumference. Initial laboratory results were consistent with severe COVID-19 pneumonia, showing elevated inflammatory markers (C-reactive protein). Arterial blood gas (ABG) analysis revealed critical hypoxemia, with a PaO₂/FiO₂ ratio of 140 in the first case, meeting the criteria for acute respiratory distress syndrome (ARDS).

This report details the clinical course of these two patients, focusing on the use of NIV as the primary ventilatory strategy. We describe the challenges encountered, including the development of pneumothorax and the unmasking of severe obstructive sleep apnea, and integrate these experiences with a review of the literature. Our aim is to provide a structured management framework for ARF in patients with Madelung's disease, emphasizing a strategy that prioritizes the avoidance of high-risk airway interventions.

Case Presentation

Case 1

A 65-year-old male with a known history of Madelung's disease presented to the emergency department with acute hypoxemic respiratory failure secondary to confirmed COVID-19 pneumonia. His body mass index (BMI) was 30 kg/m². Upon admission, his vital signs were indicative of severe distress: temperature 38.7°C, respiratory rate 37 breaths/min, heart rate 120 bpm, blood pressure 140/60 mmHg, and an oxygen saturation (SpO₂) of 82% on room air. Arterial blood gas (ABG) analysis revealed a PaO₂ of 42 mmHg and a PaO₂/FiO₂ ratio of 140, meeting criteria for acute respiratory distress syndrome (ARDS).

Given the high risk of difficult or failed intubation associated with Madelung's disease, a strategy of non-invasive ventilation (NIV) was initiated as first-line therapy. He was admitted to the intensive care unit (ICU) and placed on a bilevel positive airway pressure (BiPAP) device (V60) via a total face mask. Settings were titrated to an inspiratory positive airway pressure (IPAP) of 18 cm H₂O and an expiratory positive airway pressure (EPAP) of 8 cm H₂O to achieve adequate tidal volumes and maintain SpO₂ between 92-94%.

His ICU course was complicated by the development of bilateral pneumothorax on day 3 of NIV therapy, which was managed conservatively with thoracic drainage. Despite this complication, NIV was continued. The patient showed gradual improvement, allowing for de-escalation to high-flow nasal oxygen therapy (HFNO) by day 7, and subsequently to conventional low-flow oxygen. He was successfully discharged from the ICU to a general pneumonia ward on day 12.

Case 2

A 70-year-old male with a long-standing history of Madelung's disease (status post excision of multiple cervical lipomas) was admitted with dyspnea at rest and a dry cough. He had a significant history of heavy alcohol use (now abstinent for 6 years) and a 55 pack-year smoking history. Physical examination was notable for profound cervical dysmorphism with marked enlargement of the neck circumference. Initial SpO₂ was 88-90% on room air. He was diagnosed with severe COVID-19 pneumonia with bacterial superinfection.

Due to progressive global respiratory failure and the anticipated difficult airway, NIV was commenced using an oronasal mask (Stellar 150 device) in bilevel spontaneous-timed (ST) mode. Settings were IPAP 16 cm H₂O, EPAP 8 cm H₂O, with a backup rate. Weaning from ventilatory support proved challenging, punctuated by periods of gas exchange deterioration and significant nocturnal desaturation.

A diagnostic workup, including a cardio-respiratory sleep study performed post-acute recovery, revealed a previously undiagnosed moderate obstructive sleep apnea (OSA) with an apnea-hypopnea index (AHI) of 26 events/hour. This underlying condition was identified as a major contributor to the prolonged respiratory failure and difficult weaning [5].

The patient was subsequently established on home auto-adjusting positive airway pressure (APAP) therapy, which resulted in sustained clinical and gasometric improvement, facilitating his eventual discharge with outpatient follow-up.

Table 1: Summary of Case Report

Feature / Parameter	Case 1	Case 2	Clinical Implication / Insight
Age / Sex	65-year-old male	70-year-old male	
Primary Trigger for ARF	Severe COVID-19 pneumonia (ARDS criteria)	Severe COVID-19 pneumonia with bacterial superinfection	Highlights a common modern precipitant of ARF in vulnerable patients.
Key Comorbidity	Madelung's disease, Obesity (BMI 30)	Madelung's disease, Severe OSA (AHI=26), Heavy smoking/alcohol history	Case 2 underscores the critical link between Madelung's and undiagnosed OSA, a major factor in weaning failure.
Initial ABG (PaO ₂ /FiO ₂)	140 (Severe ARDS)	286 (Moderate ARDS)	Both cases presented with significant hypoxemia justifying ventilatory support.
Primary Ventilatory Strategy	First-Line (BiPAP via Total Face Mask) NIV	First-Line (BiPAP via Oronasal Mask)	Active avoidance of intubation due to known difficult airway. Case 1 demonstrates interface choice to prevent skin lesions.
Major Complication During NIV	Bilateral pneumothorax (Day 3)	Difficult weaning, nocturnal desaturation	Shows NIV can be sustained even with complications; also reveals underlying OSA as a cause of prolonged failure.
Key Diagnostic Finding		Formal sleep study confirming moderate OSA	This diagnosis changed long-term management (home APAP).
Outcome & Disposition	Successful weaning to HFNO, discharged from ICU on Day 12.	Successful discharge on home auto-CPAP/APAP after OSA diagnosis and treatment.	NIV was a bridge to recovery in Case 1 and a bridge to diagnosis of a chronic condition in Case 2.

Table 2: Summary Respiratory Management in Respiratory Failure (niv and IMV)

Ventilation methodology		NIV	IMV
	Indications	Respiratory distress due to a condition exacerbated by Madelung's disease	Severe respiratory failure, acute exacerbation
	Concept	Positive pressure ventilation delivered through an interface	Controlled ventilation through an endotracheal or tracheostomy tube
	Ventilatory Modes	CPAP, BiLevel	Assist-control, pressure support, synchronized intermittent mandatory ventilation
	Monitoring	Regular assessment of response, adjustment of settings, detection of complications	Monitoring of oxygenation, ventilation, arterial blood gases, and pulse oximetry
	Weaning	Transition to spontaneous breathing, extubation readiness assessment	Weaning protocols to assess readiness for extubation or transitioning to non-invasive ventilation

Table 3: Summary Lung Disorders and Approach in Madelung Disorders

Summary lung disorders in Madelung's Disease		Therapeutic Approach
Respiratory Complications	Difficulty breathing	Weight management, treatment of underlying conditions
	Sleep apnea	Continuous positive airway pressure (CPAP) therapy
	Upper airway obstruction	Surgery to remove excess adipose tissue
	Dyspnea (shortness of breath)	Weight management, respiratory physiotherapy
	Reduced lung capacity	Pulmonary rehabilitation
Changes in the Upper Airways	Narrowing of the upper airways due to compression from adipose tissue	Surgery to remove excess adipose tissue
	Snoring	Weight management, treatment of underlying causes (e.g., sleep apnea)
Impaired Lung Function	Decreased lung capacity due to lung compression	Pulmonary rehabilitation, respiratory exercises

Discussion

This case series demonstrates the feasibility and challenges of employing non-invasive ventilation (NIV) as a primary strategy for managing acute respiratory failure (ARF) in patients with Madelung's disease, a population at exceptionally high risk during emergency airway management.

NIV as a Strategic Imperative to Avoid a Hazardous Airway

The paramount concern in managing ARF in Madelung's disease is the well-documented high incidence of

difficult or failed endotracheal intubation [1,2,6]. The symmetric cervical lipomatosis distorts anatomy, limiting neck extension and obscuring laryngeal views. Our management decision to initiate NIV first-line in both cases was driven directly by this imperative. The successful outcome avoiding intubation entirely strengthens the argument that NIV should not be merely an alternative but a primary and deliberate strategy in the initial management plan for these patients, whenever clinically permissible.

Practical NIV Implementation: Interface and Vigilance

Successful NIV in this context requires careful technical execution. In Case 1, the use of a total face mask was instrumental. Patients with Madelung's disease often have irregular facial contours and sensitive skin due to underlying lipomas. The total face mask distributes pressure over a larger area, which likely contributed to the absence of skin breakdown in our patient, a complication reported with other interfaces [3]. Furthermore, clinicians must be vigilant for complications common to both NIV and the underlying condition. The development of bilateral pneumothorax in Case 1, while serious, was managed without intubation, demonstrating that certain NIV complications can be managed conservatively without abandoning the non-invasive strategy.

The Critical Role of Sleep-Disordered Breathing: Beyond Acute Management

The most significant insight from Case 2 is the elucidation of obstructive sleep apnea (OSA) as a central comorbidity shaping the clinical course. The patient's difficult weaning and nocturnal hypoventilation were initially puzzling until a formal sleep study revealed moderate OSA (AHI 26). This finding is not coincidental but a recognized association in Madelung's disease, thought to be caused by fatty infiltration compromising upper airway patency [5,7].

Our case powerfully illustrates that ARF in these patients may be an acute-on-chronic phenomenon. Therefore, the management algorithm must extend beyond treating the acute precipitant (e.g., pneumonia). We propose that routine screening for OSA, either clinically or via sleep studies upon clinical stabilization, should be integrated into the care pathway. As shown, treating the underlying OSA with home positive airway pressure therapy was transformative

for the patient's long-term respiratory status.

Synthesis and Proposed Management Framework

Based on our experience and literature synthesis, we propose a structured approach to ARF in Madelung's disease: (1) Early identification of Madelung's disease as a "difficult airway alert" upon admission; (2) A trial of NIV with a carefully selected interface (consider total face mask) as the initial strategy for moderate to severe ARF; (3) Vigilance for complications like pneumothorax; (4) Investigation for concomitant OSA in cases of prolonged weaning; and (5) Long-term planning including management of chronic respiratory issues like OSA.

Patient Perspective

Both patients expressed profound relief at avoiding invasive endotracheal intubation, a procedure they were informed carried exceptionally high risk due to their anatomical condition. For the patient in Case 2, the diagnosis of obstructive sleep apnea (OSA) was a pivotal moment. He reported that prior to this admission, his chronic fatigue and unrefreshing sleep were unexplained burdens. The initiation of home APAP therapy not only resolved his nocturnal symptoms but also provided a clear understanding of a long-standing health issue, significantly improving his overall quality of life and engagement with his long-term health management. These perspectives underscore that successful management in such complex cases extends beyond acute physiological support to include diagnosis of underlying comorbidities and patient education.

Study Limitations

Our conclusions are drawn from a limited number of cases (n=2), which is inherent to studying rare diseases. Furthermore, the specific challenge was ARF triggered by COVID-19; outcomes may vary with other etiologies. Nevertheless, the principles of airway risk mitigation and comprehensive search for contributing factors like OSA remain broadly applicable.

Conclusion

In conclusion, managing ARF in Madelung's disease requires a paradigm shift away from default intubation. A proactive NIV-first strategy, informed by an understanding of the difficult airway and a high index of suspicion for comorbid OSA, can successfully avert life-threatening procedures and guide more holistic patient care.

Conflict of Interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Author Contributions

BG and AE conceived the study. FA and BG collected and analyzed the clinical data. FA wrote the first draft of the manuscript. All authors (BG, AE, FA) contributed to the critical revision of the manuscript, approved the final version, and agree to be accountable for all aspects of the work.

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Data Availability Statement

The original clinical data presented in this case report are included within the article. No new datasets were generated or analyzed.

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