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Ventilatory Management Issues in Acute Respiratory Failure Among Patients with Amyotrophic Lateral Sclerosis

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ABSTRACT

Introduction: Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disease leading to impairment of the respiratory muscles, which causes respiratory failure, the primary cause of death in these patients. Management of acute respiratory distress in ALS patients is particularly challenging due to the absence of a curative treatment. Invasive ventilation may prolong survival but does not alter the inevitable course of the disease.

Case Reports: We report two cases of patients with ALS admitted to the emergency department for acute respiratory failure.

The first patient, a 36-year-old male, developed acute respiratory distress secondary to aspiration. Upon admission, his SpO_2 was below 70%, blood pressure 110/70 mmHg, heart rate 110 bpm, respiratory rate 30 breaths/min, with diffuse bronchial crackles on pulmonary auscultation. He underwent intubation followed by invasive mechanical ventilation and ultimately required tracheostomy due to ventilator dependency. His condition deteriorated, and he died three months after hospitalization.

The second patient, aged 42, developed Legionella pneumonia complicated by acute respiratory failure. He presented with SpO_2 below 80%, blood pressure 110/60 mmHg, heart rate 100 bpm, respiratory rate 26 breaths/min, and bronchial congestion. He also required intubation followed by tracheostomy. Unlike the first case, he remains alive under assisted ventilation but is in a state of complete dependency.

Conclusion: These cases illustrate the complexity of emergency care in ALS patients with acute respiratory failure. Although intubation and invasive ventilation are sometimes unavoidable, they often result in prolonged dependency, raising questions about their benefit-risk ratio. In countries where guidelines exist, decisions are made in accordance with the patient's advance directives and through multidisciplinary deliberation. However, in our context, these principles are rarely followed, leading to often improvised emergency therapeutic decisions. Close collaboration between emergency physicians and neurologists is essential to optimize management and avoid prolonged invasive interventions that may not align with the patient's prognosis.

Keywords: Amyotrophic lateral sclerosis, respiratory distress, emergency care, invasive ventilation, medical ethics.

INTRODUCTION

Amyotrophic lateral sclerosis (ALS), also known as Charcot disease, is a progressive neurodegenerative disorder affecting motor neurons, with an annual incidence of 1.5 to

2.5 cases per 100,000 inhabitants and a prevalence of 6 to 8 per 100,000, primarily affecting individuals over 40 years old. The median survival time is 30 months after diagnosis, but less than 6 months once diaphragmatic involvement occurs. There is no curative treatment, although riluzole (Rilutek®) and edaravone (Radicava®) offer modest survival benefits.

To optimize ALS management, the literature emphasizes the effectiveness of a multidisciplinary approach. A study by Van den Berg et al. (2005) showed that patients followed in multidisciplinary centers—including neurologists, pulmonologists, physiotherapists, and nutritionists—have an average survival prolongation of 7.5 months compared to conventional care [1]. This approach enables early detection of complications and rapid adaptation of care. Within this framework, another key pillar is non-invasive ventilation (NIV). According to Bourke et al. (2006), published in *The Lancet Neurology*, NIV extends median survival by 11 months in ALS patients without severe bulbar involvement ($p < 0.001$) [2], while a meta-analysis by Piepers et al. (2006), in the *Journal of Neurology*, confirmed that NIV reduces hospitalization rates by 30% by stabilizing respiratory function at an early stage [3].

However, despite these advances, a major challenge arises with invasive ventilation, sometimes unavoidable in the face of acute respiratory failure (ARF). Although effective in the short term, this option often results in prolonged dependency. Sancho et al. (2018) reported that 60% of patients on invasive ventilation become long-term dependent, raising ethical and practical dilemmas [4]. These challenges are particularly complex given the frequent absence of advance directives and clear protocols.

Through the analysis of two clinical cases, this article explores the challenges of ventilatory management of acute respiratory failure in the emergency setting for patients with amyotrophic lateral sclerosis.

PATIENT AND OBSERVATION

Case 1:

This was a 36-year-old patient diagnosed with ALS for 20 months, who presented to the emergency department with acute respiratory failure caused by Aspiration of gastric contents.

Upon arrival at the emergency room, vital signs were: temperature 37.2 °C, blood pressure 110/70 mmHg, heart rate 110 bpm, respiratory rate 30 breaths/min, and SpO_2 70% on room air. Physical examination revealed normal alertness with bronchial rales on lung auscultation. The remainder of the clinical examination was normal. Initial laboratory tests showed severe respiratory acidosis (pH 7.17, PCO_2 56 mmHg, bicarbonates 22 mmol/L). The patient was initially treated with non-invasive ventilation (NIV), but its failure led to invasive mechanical ventilation (IMV), followed by tracheostomy on day 10 due to persistent respiratory dependence. Despite these interventions, His condition deteriorated, and he died three months after hospitalization.

Case 2:

This involved a 42-year-old man diagnosed with ALS for 26 months, who developed Legionella pneumophila pneumonia complicated by acute respiratory distress. Clinically, the patient was mildly confused. There were no neurological localization signs; pupils were slightly dilated but reactive, and diffuse crackles were noted on lung auscultation. Vital signs were: blood pressure 110/60 mmHg, heart rate 100 bpm, respiratory rate 26 breaths/min, oxygen saturation below 80% on room air, and temperature 38.8 °C. Laboratory tests showed leukocytosis at 14,000 cells/mm³, hemoglobin at 11 g/dL, platelets at 241,000/mm³, and respiratory acidosis on blood gas analysis (pH 7.22, PCO_2 54 mmHg, bicarbonates 24 mmol/L). NIV was attempted unsuccessfully, followed by invasive mechanical ventilation within the hour and tracheostomy on day 14. Unlike the first case, this patient is currently alive

but fully dependent on assisted ventilation, with a severely compromised quality of life.

DISCUSSION

Amyotrophic lateral sclerosis (ALS) is an incurable disease characterized by progressive degeneration of both upper and lower motor neurons, leading to generalized muscle weakness. As highlighted by Hardiman et al., respiratory muscle weakness is a common feature of ALS, typically occurring in advanced stages of the disease and leading to acute respiratory failure (ARF), which is the leading cause of death [5]. The clinical cases presented illustrate this trajectory: ARF occurred at 20 and 26 months after diagnosis, a timeline consistent with the natural progression of the disease [7]. The precipitating factors observed—aspiration in the first case and Legionella pneumophila pneumonia in the second—reflect typical respiratory complications. Indeed, Lechtzin et al. reported that the most frequent causes of emergency admissions were pneumonia (34%) and aspiration events (11%) [8]. These events reveal a critical respiratory vulnerability related to diaphragmatic and intercostal muscle weakness, severely compromising patients' ability to cope with acute insults [9].

In response to this respiratory vulnerability, non-invasive ventilation (NIV) is widely recommended as first-line therapy in the management of ALS patients due to its proven efficacy in improving quality of life and prolonging survival [2]. However, the two clinical cases reported here highlight NIV failure, which can be explained by several mechanisms:

- The advanced disease stage, at 20 and 26 months, suggests respiratory muscle weakness too severe for NIV to adequately compensate for ARF.
- The absence of optimized prior follow-up, including early initiation of NIV, as noted by Radunovic et al., who emphasize that "NIV is more beneficial when started early, before significant respiratory deterioration" [11].
- Finally, NIV proves inadequate in complex acute contexts, such as those observed in the two reported cases, where complications increase ventilatory demands beyond the capacities of NIV, which is primarily designed to support chronic ventilation rather than manage severe acute crises [9].

When NIV fails, invasive mechanical ventilation (IMV) becomes an option, but it raises major ethical dilemmas related to balancing survival prolongation and quality of life preservation. Literature data converge to assert that there is no clear evidence that IMV improves long-term survival, and its use must be carefully weighed against risks of prolonged dependence [6,10,11], highlighting the limits of this approach in the face of the inexorable progression of the disease. These findings are consistent with the clinical cases presented, which concretely illustrate the challenges and constraints emphasized by the authors. In the first case, death occurred on day 12 despite IMV and a tracheostomy performed on day 10, calling into question the usefulness of such invasive interventions at an advanced stage. Conversely, in the second case, the patient survives under IMV but is fully dependent with a severely compromised quality of life. These situations reveal a dilemma between the medical goal of sustaining life and the ethical imperative to avoid unnecessary suffering. To resolve this conflict, McCluskey et al. stress the importance of advance care planning (ACP), a process that helps patients understand and share their personal values, life goals, and preferences regarding future medical care [12]. This approach provides a framework to guide therapeutic choices, particularly in advanced disease stages where patients must understand that ALS continues to progress despite respiratory support.

These therapeutic and ethical choices are even more complex in a local context that amplifies the inherent challenges of ALS.

Our environment presents several aggravating specifics:

- A lack of prognosis awareness by patients and their families, often due to insufficient communication at diagnosis, limits their involvement in informed decision-making, a difficulty also noted in the literature [12].
- The notable absence of palliative care structures deprives patients of an approach focused on comfort and dignity, recognized as crucial at end-of-life, leaving medical teams helpless in the face of ethical dilemmas [13]. Oliver et al. specify that "early integration of palliative care improves quality of life and reduces unnecessary hospitalizations" [14], a benefit currently out of reach due to lack of local resources.
- The lack of clear protocols and interdisciplinary collaboration among neurologists, pulmonologists, and intensivists, essential for optimal management, forces caregivers to improvise under pressure, turning a medically complex situation into a systemic challenge where therapeutic choices, such as transition to IMV, are more reactive than the result of a thoughtful, coordinated strategy [10]. According to Simmons, teamwork and consultation within structured multidisciplinary care models are indispensable to ensure optimal and proactive management of ALS [13].

All these observations underline the problematic and difficulties of managing ALS patients in emergency settings, where challenges extend beyond medical aspects to include organizational, ethical, and psychosocial dimensions, requiring adapted responses

CONCLUSION

The management of acute respiratory failure (ARF) in patients with amyotrophic lateral sclerosis (ALS) highlights major clinical and ethical challenges, exacerbated by local structural shortcomings. Non-invasive ventilation (NIV), although effective in early stages, shows its limitations in advanced disease or acute crises, while invasive mechanical ventilation (IMV) raises ethical dilemmas related to dependence and quality of life, without guaranteeing significant prolonged survival. Our work, illustrated by the two clinical cases, reveals several shortcomings in this context. In light of these issues, several recommendations are warranted:

- Enhanced communication from the time of diagnosis is essential to inform patients and their families, promoting advance care planning aligned with their values.
- The establishment of standardized local protocols, incorporating a multidisciplinary approach among neurologists, pulmonologists, and intensivists, would reduce hasty interventions and optimize therapeutic decisions.
- The development of palliative care facilities would improve end-of-life quality and support medical teams facing ethical dilemmas.
- Psychosocial aspects, completely absent in our setting, constitute a critical gap; integrating psychologists into patient care becomes imperative to address this void and meet the essential needs of patients and their families

Declaration by Authors

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