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ANESTHETIC MANAGEMENT OF A PATIENT WITH AMYOTROPHIC LATERAL SCLEROSIS UNDERGOING FLEXIBLE URETERORENOLITHOTRIPSY: A CASE REPORT

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ABSTRACT

Introduction: Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disease characterized by motor weakness and respiratory impairment, increasing perioperative risk, particularly during anesthetic induction, ventilation, and extubation. **Case Report:** A 62-year-old female patient, 83 kg, ASA III, with a five-year history of ALS, an ALSFRS-R score of 10, and dependence on bilevel noninvasive ventilation (BiPAP), underwent flexible ureterorenolithotripsy with double-J stent placement. Preoperative evaluation revealed severe functional impairment and high respiratory risk. Total intravenous anesthesia with propofol and remifentanil using target-controlled infusion was performed without neuromuscular blocking agents, combined with bispectral index and train-of-four monitoring. Orotracheal intubation was successfully achieved using videolaryngoscopy (Cormack-Lehane grade I), followed by protective mechanical ventilation. The intraoperative course was uneventful, with hemodynamic stability and adequate oxygenation. At the end of the procedure, careful extubation was performed with immediate reconnection to noninvasive ventilation, and the patient was transferred to the post-anesthesia care unit under continuous monitoring without adverse events. **Discussion:** Anesthetic management in patients with ALS requires an individualized approach, considering reduced ventilatory reserve and unpredictable responses to neuromuscular blocking agents, highlighting the importance of strategies that preserve spontaneous ventilation and minimize respiratory depression. In this context, avoidance of neuromuscular blockers, combined with depth of anesthesia and neuromuscular monitoring, as well as early postoperative ventilatory support, proved effective in reducing the risk of respiratory complications. **Conclusion:** Risk anticipation, careful anesthetic titration, and immediate postoperative ventilatory planning are essential to optimize anesthetic safety in patients with advanced ALS.

Keywords: Amyotrophic lateral sclerosis, General anesthesia, Neuromuscular blocking agents, Noninvasive ventilation, Perioperative period.

INTRODUCTION

Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disease that affects upper and lower motor neurons, leading to progressive muscle weakness, bulbar dysfunction, and respiratory impairment over the course of the disease.[1]

Respiratory failure is the leading cause of morbidity and mortality in these patients, often already present in more advanced stages, with the need for home noninvasive ventilation.[2] In addition, bulbar dysfunction increases the risk of pulmonary aspiration, while respiratory muscle weakness impairs both spontaneous ventilation and postoperative weaning. In other words, even procedures considered minor carry a significantly higher risk compared with the general population.[3]

In the anesthetic context, these patients exhibit a variable response to neuromuscular blocking agents, with a risk of hyperkalemia associated with the use of succinylcholine and increased sensitivity to nondepolarizing agents.[1,3] In addition, there is greater vulnerability to respiratory depression induced by anesthetics and opioids, requiring cautious management and, whenever possible, strategies that preserve spontaneous ventilation.[2,4]

Therefore, anesthetic management should be carefully planned at all stages, including induction, maintenance, extubation, and ventilatory support in the immediate postoperative period, considering the high risk of extubation failure and reintubation in these patients.[2,4]

In view of this, the present report aims to describe the anesthetic management of a patient with advanced ALS undergoing flexible ureterorenolithotripsy, highlighting clinical decisions aimed at reducing respiratory and hemodynamic complications, with a focus on a safer perioperative period.

CASE REPORT

A 62-year-old female patient underwent flexible ureterorenolithotripsy with placement of a double-J catheter on January 12, 2026, at Hospital do Rim (HR). At the pre-anesthetic evaluation, she weighed 83 kg, was 185 cm tall, and had fasted for more than 8 hours for solids and more than 3 hours for liquids. She reported a diagnosis of ALS approximately five years earlier, with progressive muscle weakness below the neck and dependence on home noninvasive bilevel positive airway pressure ventilation ("Bilevel"), findings consistent with advanced stages of the disease. She was on continuous treatment with metformin, gabapentin 400 mg, and riluzole 50 mg, and reported an allergy to pregabalin. A history of previous surgical procedures was reported, although without detailed description.

Functional assessment was performed using the "Revised Amyotrophic Lateral Sclerosis Functional Rating Scale" (ALSFRS-R), an instrument widely used to assess the functional progression of ALS. The scale consists of 12 items, each scored from 0 to 4, resulting in a total score ranging from 0 to 48 points, with higher values indicating better functional capacity. The patient had a total score of 10 points, indicating severe functional impairment and a high perioperative respiratory risk. Observational studies have shown that patients with scores below 20 are at greater risk of respiratory failure and have a worse short-term prognosis.

On physical examination, she had good dental condition but reduced cervical mobility. She was classified as ASA III.

The patient was admitted to the operating room while using Bilevel, and preoxygenation with

positive pressure was performed. Total intravenous anesthesia with propofol and remifentanil in a target-controlled infusion system was chosen, a strategy that allowed continuous anesthetic titration and performance of the procedure without the need for neuromuscular blocking agents. Monitoring included continuous electrocardiography, pulse oximetry, noninvasive blood pressure, temperature monitoring, bispectral index (BIS), and neuromuscular monitoring with train-of-four (TOF), in accordance with recommendations for patients with neuromuscular disorders. Anesthetic induction was performed with propofol and remifentanil in a target-controlled infusion system, without the use of neuromuscular blocking agents. Orotracheal intubation was performed with the aid of a videolaryngoscope, with a Cormack-Lehane grade I view, using an 8.0 cuffed orotracheal tube. Tube position was confirmed by capnography and bilateral pulmonary auscultation. Anesthesia was maintained with propofol and remifentanil, with BIS values maintained between 30 and 40.

During the intraoperative period, the patient remained hemodynamically stable with adequate oxygenation. Ceftriaxone was administered for antibiotic prophylaxis, dipyrone for analgesia, and ondansetron and dexamethasone for prophylaxis of nausea and vomiting. At the end of the procedure, extubation was performed without complications, with immediate reconnection to Bilevel for ventilatory support, an approach recommended to reduce the risk of postoperative respiratory failure in patients with ALS. The patient was transferred to the post-anesthesia care unit in a hemodynamically stable condition.

DISCUSSION

Anesthetic management in patients with ALS should primarily consider the degree of functional and respiratory impairment, since reduced ventilatory reserve significantly limits the margin of safety in the perioperative period.[1,4] In this context, respiratory muscle involvement, which is often underestimated during clinical assessment, contributes to greater susceptibility to respiratory failure following anesthetic interventions, even in minor procedures.[2,4]

Given this physiological limitation, the choice of anesthetic technique should prioritize strategies that reduce respiratory depression, allow predictable recovery, and minimize factors associated with postoperative ventilatory failure.[1,4] In this regard, total intravenous anesthesia with propofol and remifentanil, as used in the present case, allowed adequate control of anesthetic depth, while also enabling the procedure to be performed without the need for neuromuscular blocking agents, which represents a relevant advantage in this patient profile.

The use of neuromuscular blocking agents, in turn, constitutes a critical aspect of anesthetic management in ALS. Succinylcholine should be avoided due to the risk of hyperkalemia resulting from upregulation of extrajunctional nicotinic receptors.[3] Additionally, nondepolarizing neuromuscular blocking agents may have a prolonged effect and an unpredictable response, particularly in advanced stages of the disease.[2] Therefore, whenever possible, avoidance of neuromuscular blockade should be considered as a safety strategy.

When the use of neuromuscular blocking agents becomes necessary, quantitative monitoring with TOF plays a fundamental role, allowing objective assessment of the degree of blockade and reducing the risk of residual blockade in the postoperative period.[2,3] This approach is particularly relevant in patients with neuromuscular diseases, in whom small pharmacodynamic variations may result in significant clinical impact.

Figure 2. (A) Patient positioning for pre-oxygenation. (B) Video laryngoscopy.

Another determining factor in the outcomes of these patients is extubation management, which should be performed carefully, considering not only the level of consciousness but also ventilatory capacity, muscle strength, and adequate airway protection.[2,4] In this setting, the early use of noninvasive ventilation in the postoperative period, as observed in the present case, constitutes an important strategy for supporting the respiratory muscles and reducing the risk of ventilatory decompensation.[4]

Thus, successful anesthetic management in patients with ALS depends on the integration of knowledge of the pathophysiology of the disease, detailed functional assessment, and individualized decision-making, with a focus on preventing respiratory complications and optimizing postoperative recovery.

CONCLUSION

Anesthetic management in patients with advanced ALS requires an individualized approach, with emphasis on preserving respiratory function and reducing perioperative complications. In the present case, the use of total intravenous anesthesia without neuromuscular blocking agents, combined with careful extubation and immediate reconnection to noninvasive ventilation, proved to be an effective strategy for maintaining ventilatory stability in the immediate postoperative period, reinforcing that anticipation of respiratory complications, appropriate titration of anesthetic agents, and planning of postoperative ventilatory support are essential measures to optimize anesthetic safety in these patients.

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EDITORIAL AND REVIEW

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