Open Access Original Research Article

MedPeer Publisher
Abbreviated Key Title: MedPeer
ISSN : 3066-2737
homepage: https://www.medpeerpublishers.com

KEYS TO SUCCESSFUL ANESTHETIC MANAGEMENT IN 3A SYNDROME

DOI: 10.70780/medpeer.000QGQ2

AUTHOR AND AFFILIATION

Hamza Kassimi [1], Larbi Ed-dafali [2], Alae Alae El koraichi [2] , Salma Ech Cherif El Kettani [2], Aziza Bentalha [2]
[1] Anesthesiology Departement, Mohammed 5 Military Hospital of Rabat, MAROC
[2] Anaesthesiology department, Children Hospital of Rabat, MAROC

Corresponding author: Hamza Kassimi.

ABSTRACT

Allgrove syndrome, also known as triple A syndrome, is a rare genetic disorder characterized by adrenal insufficiency, achalasia, and alacrima, with potential neurological and autonomic complications. This complex clinical triad poses a significant anesthetic challenge, requiring meticulous preparation that integrates the pathophysiological specificities of the disease to ensure perioperative patient safety.
We report the case of a 9-year-old patient with Allgrove syndrome who underwent Heller's myotomy for refractory achalasia. Perioperative management included hydrocortisone replacement therapy, rapid sequence induction to reduce the risk of pulmonary aspiration, strict glycemic monitoring, and ocular protection to prevent complications related to alacrima.

KEYWORDS

adrenal insufficiency, achalasia, perioperative management, triple a syndrome, allgrove syndrome

MAIN ARTICLE

Introduction

Allgrove syndrome, also known as 3A syndrome, is a rare autosomal recessive genetic disorder first described in 1978 by Allgrove et al. [1]. It is characterized by the triad of adrenal insufficiency, achalasia of the esophagogastric junction, and alacrima, with additional neurological and autonomic abnormalities [2, 3].

The responsible gene, AAAS, located on chromosome 12q13, encodes the ALADIN protein, which is essential for proper nuclear pore function [4]. Mutations in this gene disrupt various cellular processes, predisposing patients to systemic complications, particularly during periods of physiological or surgical stress.

The combination of metabolic and organ dysfunction poses significant anesthetic challenges. Adrenal insufficiency requires meticulous glucocorticoid substitution to prevent acute adrenal crises [5]. Achalasia increases the risk of pulmonary aspiration [6], and autonomic dysfunction can lead to unpredictable hemodynamic instability [7]. These challenges necessitate a multidisciplinary approach and careful preoperative planning.

This report describes the anesthetic management of a 9-year-old girl with Allgrove syndrome undergoing Heller's cardiomyotomy for refractory achalasia. This case highlights the perioperative management nuances of this rare condition and outlines strategies to optimize anesthetic safety.

Case Presentation

The patient, a 9-year-old girl weighing 24 kg, was born to non-consanguineous parents and was diagnosed with Allgrove syndrome at the age of 5. The diagnosis was based on the triad of alacrima, achalasia confirmed by esophageal manometry, and adrenal insufficiency documented biochemically. Additionally, electromyography revealed moderate axonal neuropathy. While on hydrocortisone and artificial tears, the patient underwent two pneumatic esophageal dilations under general anesthesia for symptomatic achalasia. Due to persistent dysphagia (to solids and liquids) and recurrent postprandial vomiting, Heller's cardiomyotomy was indicated (figure *1*).

During the pre-anesthetic evaluation, clinical examination revealed growth retardation (-1 SD for height and -2 SD for weight), good venous access, normal cardiopulmonary auscultation, and no signs of neurovegetative dysautonomia on postural testing. Preoperative laboratory

tests showed no abnormalities, with normal serum sodium, potassium, and glucose levels. The patient received 10 mg of oral prednisolone the day before surgery, and a six-hour fasting period was observed.

In the operating room, a peripheral venous line (22G) was already in place. Standard monitoring included electrocardiography, pulse oximetry, and non-invasive blood pressure measurement. An arterial line was deemed unnecessary due to hemodynamic stability. Preoperative fluid replacement with 0.9% saline corrected a fasting-related fluid deficit of 384 mL, administered at an initial infusion rate of 240 mL/h for the first hour, followed by 144 mL/h thereafter. A gastric tube was placed and aspirated before induction. Rapid sequence induction was performed using propofol (90 mg), rocuronium (15 mg), and fentanyl (60 µg). Orotracheal intubation was successfully achieved using a 5.5-mm cuffed tube. Ocular protection included eye drops and compresses. Hydrocortisone (60 mg) was administered intravenously. Maintenance anesthesia was achieved with sevoflurane, with ventilatory settings adjusted to maintain end-tidal CO2 between 35 and 45 mmHg . A second peripheral venous line (22G) was placed after induction. Capillary blood glucose was monitored every 30 minutes and remained stable between 0.8 and 1 g/dL throughout the procedure.

Postoperative analgesia was managed with paracetamol (360 mg). A continuous hydrocortisone infusion (60 mg/day) was maintained for 48 hours, followed by gradual tapering to the maintenance dose. No electrolyte imbalances or ocular or neurological complications were observed

Discussion

The anesthetic management of this patient presented several challenges related to the pathophysiology of Allgrove syndrome and its clinical manifestations. The definitive diagnosis, established at the age of 5, was based on the classic triad of alacrima, achalasia, and adrenal insufficiency [1, 2]. Axonal neuropathy, identified by electromyography, is a common complication that can impair autonomic responses [8]. Thorough preoperative evaluation revealed growth retardation, a typical finding in these patients [9]. The stable clinical condition and absence of neurovegetative dysautonomia guided the anesthetic approach. Prophylactic prednisolone administration was employed to prevent adrenal crises, a strategy supported by the literature [10]. Rapid sequence induction was chosen to minimize the risk of pulmonary aspiration associated with achalasia [11, 12]. The use of propofol, rocuronium, and fentanyl facilitated a rapid and safe orotracheal intubation,

consistent with recommendations for these patients [13]. Sevoflurane was used for maintenance anesthesia due to its stable profile and limited cardiovascular side effects [14]. Preoperative hydration with isotonic saline was essential to correct the fasting-related fluid deficit.

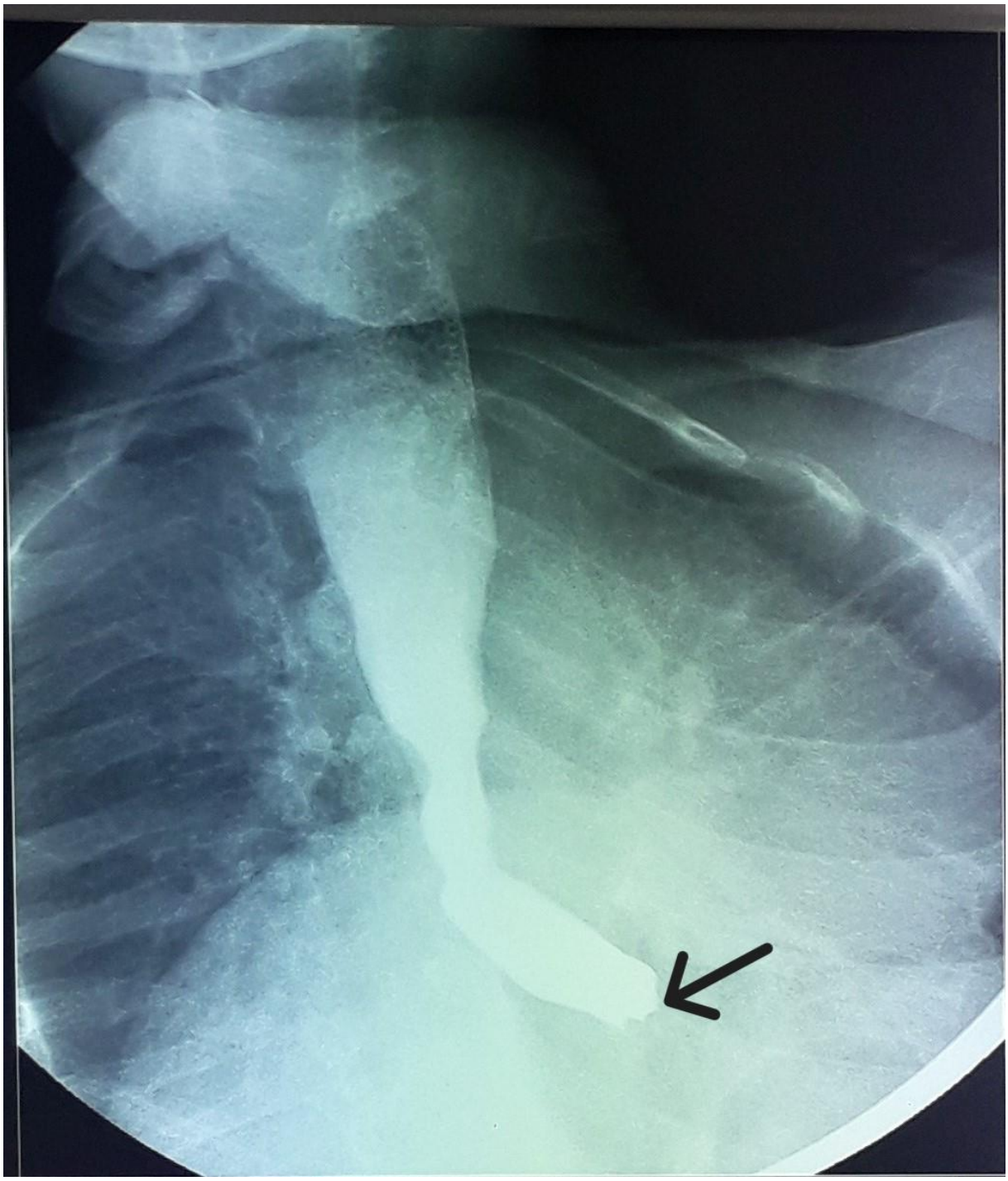
Special attention was given to glycemic control, given the increased risk of hypoglycemia in these patients [15]. Regular glucose monitoring prevented any fluctuations. Prophylactic ocular protection was necessary to prevent complications associated with alacrima [16]. Postoperatively, continuous hydrocortisone administration was initiated, followed by a gradual return to the maintenance dose [17].

Conclusion

This case highlights the complexity of anesthetic management in Allgrove syndrome. Precise planning, including rigorous hormonal substitution, rapid sequence induction for full-stomach precautions, strict glycemic monitoring, and ocular protection, is essential to minimize complications. A multidisciplinary team approach is a significant asset in ensuring perioperative safety and efficacy.

FIGURES:

Figure 1: Upper gastrointestinal series showing significant esophageal dilatation proximal to the stenosis at the lower esophageal sphincter, as indicated by the arrow



ACKNOWLEDGEMENTS

The authors have no acknowledgements to declare and report no conflicts of interest.

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