Pediatric Anesthesia and Critical Care Journal 2025;13(2):51-53
doi:10.14587/paccj.2025.8

PACCJ

Anesthetic management of percutaneous nephrolithotripsy in an 11 year old girl with Rett Syndrome: a case report

R. N. Naghiyev, F. A. Ahmadov, E. M. Nasibova

Azerbaijan Medical University, Baku, Azerbaijan

Corresponding author: E. M. Nasibova, Azerbaijan Medical University. Email: doc.nasibova.esmira@gmail.com

Keypoints

This case illustrates that sevoflurane-based anesthesia combined with caudal block, without the use of opioids or neuromuscular blockers, is safe and effective for children with Rett Syndrome.

Abstract

Background: Rett syndrome (RTT) is a rare X-linked neurodevelopmental disorder primarily affecting females, characterized by severe cognitive impairment, autonomic dysregulation, epilepsy, and abnormal respiratory patterns. Anesthetic management in these patients is particularly challenging due to increased sensitivity to medications, airway instability, and potential for postoperative complications. We report the anesthetic management of an 11-year-old girl (15 kg) with Rett syndrome undergoing elective left-sided percutaneous nephrolithotripsy for staghorn calculus. Premedication was administered with oral midazolam (0.5 mg/kg) mixed in cherry juice. Stepwise inhalational induction with sevoflurane was performed, followed by successful orotracheal intubation without neuromuscular blockade. A single-shot caudal block with bupivacaine 0.5% (50 mg), dexamethasone 4 mg, and normal saline was performed. No intraoperative opioids or muscle relaxants were used. Intraoperative hemodynamics and postoperative recovery were stable. This case illustrates that sevoflurane-based anesthesia combined with caudal block, without the use of opioids or neuromuscular blockers, is safe and effective for children with RTT undergoing urological surgery. Regional anesthesia techniques can enhance perioperative stability in patients with complex neurodevelopmental disorders.

Keywords

Rett syndrome, pediatric anesthesia, caudal block, sevoflurane, neurodevelopmental disorders, case report

Introduction

Rett syndrome (RTT) is a severe neurodevelopmental disorder typically affecting females, resulting from mutations in the MECP2 gene. The condition is associated with epilepsy, irregular breathing patterns, autonomic nervous system dysfunction, and heightened sensitivity to anesthetic agents. These characteristics present significant anesthetic challenges. Careful preoperative assessment, avoidance of medications that may exacerbate respiratory or autonomic instability, and the use of regional techniques are essential components of safe anesthetic management. [1-4]

Case Report

An 11-year-old female patient (weight 15 kg) with a diagnosis of staghorn calculus of the left kidney and a confirmed clinical diagnosis of Rett syndrome was scheduled for elective left-sided percutaneous nephrolithotripsy. The patient had a history of seizures, non-verbal communication, and poor motor coordination, but no tracheostomy or feeding tube. Premedication included oral midazolam at a dose of 0.5 mg/kg (7.5 mg total) administered in cherry juice 30 minutes before the procedure. Upon

arrival in the operating room, she was in a sedated but responsive state. IV access was established, and anesthesia was induced via stepwise inhalational administration of sevoflurane. After achieving adequate depth of anesthesia, orotracheal intubation was successfully performed using a size 5 endotracheal tube without the use of neuromuscular blocking agents. The patient was positioned in the left lateral decubitus position, and a single-shot caudal block was performed using a mixture of 10 ml 0.5% bupivacaine (50 mg), 4 mg dexamethasone, and 15 ml of normal saline (total volume 29 ml). Anesthesia was maintained with sevoflurane throughout the procedure. Hemodynamic parameters remained stable throughout: blood pressure 85/50 mmHg, heart rate 82–84 bpm, SpO_2 99%. No intraoperative opioids or muscle relaxants were administered. The procedure was completed uneventfully. The patient was extubated in the operating room and transferred to the recovery area in stable condition. Postoperative recovery was smooth without respiratory or hemodynamic complications.

Discussion

Children with Rett syndrome present unique challenges for anesthetic management due to their neurological and autonomic dysfunction. Common issues include abnormal respiratory patterns such as breath-holding, hyperventilation, and apnea, increased seizure susceptibility, and muscle hypotonia. Careful avoidance of agents that depress respiration or prolong recovery is paramount. In this case, sevoflurane was chosen for induction and maintenance due to its favorable pharmacokinetics, minimal airway irritation, and rapid recovery profile. Avoidance of neuromuscular blockers minimized the risk of prolonged paralysis and unpredictable responses. The use of a caudal block provided effective intraoperative analgesia, reducing the need for systemic opioids and contributing to hemodynamic stability. The addition of dexamethasone may have prolonged the analgesic effect of the caudal block and helped reduce postoperative inflammation. Overall, this case demonstrates that with proper planning and a multimodal approach, safe anesthesia can be provided even in complex neurodevelopmental syndromes like Rett syndrome.

Conclusion

In pediatric patients with Rett syndrome undergoing surgical procedures, a tailored anesthetic plan involving sevoflurane-based inhalational anesthesia and caudal regional block without opioids or muscle relaxants can provide effective and safe perioperative management. This case supports the value of regional anesthesia techniques in minimizing drug exposure and maintaining physiological stability in patients with neurodevelopmental disorders.

References

1. Coleman P. Rett syndrome: anaesthesia management. Paediatr Anaesth. 2003 Feb;13(2):180; author reply 180.
2. Khalil SN, Hanna E, Farag A, Armendartz G. Rett syndrome: anaesthesia management. Paediatr Anaesth. 2002 May;12(4):375.
3. Som A, Baidya DK, Arora MK, Maitra S, Gupta S. Rett syndrome: a concern for the anesthesiologists. J Clin Anesth. 2016 Jun;31:247-8.
4. Arun Kumar H, Chaudhuri S, Budania LS, Joseph TT. Peculiar breathing in Rett syndrome: Anesthesiologist's nightmare. J Anaesthesiol Clin Pharmacol. 2013 Apr;29(2):278-80.