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Case Report

Anesthetic Considerations in a Pediatric Patient with Lennox-Gastaut Syndrome Undergoing Surgery: A Case Report

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Abstract: Lennox–Gastaut syndrome (LGS) is a severe developmental epileptic encephalopathy that presents anesthetic challenges because of refractory epilepsy, multiple antiepileptic medications. We report the anesthetic management of a 7-year-old boy with LGS who underwent elective tracheostomy for progressive upper airway obstruction. Preoperative evaluation revealed micrognathia, retrognathia, and dynamic upper airway obstruction, suggesting a potentially difficult airway. Antiepileptic medications were continued throughout the perioperative period, and general anesthesia was induced after preoxygenation with a HFNC (high-flow nasal cannula). Tracheal intubation was successfully performed using a McGrath MAC video laryngoscope, and anesthesia was maintained uneventfully with sevoflurane and remifentanil without perioperative seizure-related or airway complications. The patient recovered without anesthetic complications. This case highlights the importance of careful perioperative planning, including appropriate seizure management and airway preparation, for the safe administration of general anesthesia in pediatric patients with LGS.

Keywords: Lennox–Gastaut Syndrome, Epilepsy, Anesthesia, Tracheostomy, Retrognathia, High Flow Nasal Cannula.

INTRODUCTION

Lennox-Gastaut syndrome (LGS) is a rare and severe developmental epileptic encephalopathy characterized by multiple seizure types, cognitive impairment, and characteristic electroencephalographic findings of slow spike-and-wave discharges [1, 2]. The syndrome typically begins in early childhood and is often refractory to antiepileptic medications, with many patients experiencing persistent seizures throughout adulthood. Because of its chronic neurological manifestations and frequent association with developmental delay, LGS poses significant challenges in the long-term medical and surgical management of affected individuals.

Anesthetic management of patients with LGS requires careful perioperative planning because seizure control may be affected by anesthetic agents, perioperative stress, and interactions with antiepileptic medications. Furthermore, associated craniofacial abnormalities, neuromuscular dysfunction, or other comorbidities may contribute to airway management difficulties in some patients.

Herein, we report the successful anesthetic management of a patient with Lennox-Gastaut syndrome with anticipated difficult airway. Careful preparation enabled successful tracheal intubation without complications in patients with LGS.

CASE REPORT

A 7-year-old boy (height, 117 cm; weight, 21 kg) with Lennox–Gastaut syndrome (LGS) was scheduled for elective tracheostomy because of progressive upper airway hypotonia, recurrent episodes of respiratory distress, and recurrent pneumonia associated with copious purulent sputum.

The patient was born at 40 weeks and 2 days of gestation by elective Cesarean section with a birth weight

of 3.547 kg. There was no documented history of perinatal hypoxic injury or birth trauma. He had exhibited global developmental delay since infancy. At 1 year of age, he developed polymorphic seizures, including tonic, atonic, and atypical absence seizures, accompanied by regression of motor milestones. Electroencephalography (EEG) demonstrated multifocal interictal epileptiform discharges, atypical generalized spike-and-wave discharges, increased epileptic activation during sleep, and burst-attenuation patterns. The severely disorganized background activity and impaired sleep architecture were consistent with epileptic encephalopathy, and the overall clinical and electrophysiological findings supported the diagnosis of LGS. Despite treatment with multiple antiepileptic medications, including pregabalin, valproate, and zonisamide, his seizures remained poorly controlled.

Neurological examination revealed severe cognitive impairment, generalized hypotonia, and stereotypic hand movements. Brain magnetic resonance imaging demonstrated diffuse cerebral volume loss and ventriculomegaly. EEG showed diffuse frontotemporal slow spike-wave discharges. Physical examination revealed congenital craniofacial abnormalities, including micrognathia and retrognathia, raising concern for a potentially difficult airway. The patient was classified as American Society of Anesthesiologists (ASA) Physical Status IV. His regular antiepileptic medications (pregabalin, valproate, and zonisamide) were continued, and the scheduled morning doses were administered on the day of surgery to minimize the risk of perioperative seizure exacerbation.

Preoperative flexible fiberoptic laryngoscopy demonstrated a markedly reduced anteroposterior diameter of the upper airway. During inspiration, the epiglottis repeatedly contacted the posterior pharyngeal wall, resulting in dynamic upper airway obstruction. Based on these findings, difficult mask ventilation and tracheal intubation were anticipated.

Following standard monitoring, preoxygenation was performed using a HFNC for 10 minutes, improving peripheral oxygen saturation from 85% to 100%. A double-set-up strategy was adopted, with the surgical team prepared to perform an emergency cricothyrotomy if airway management failed. General anesthesia was induced with intravenous propofol (40 mg) and rocuronium (20 mg). Tracheal intubation was successfully achieved on the first attempt using a McGrath MAC video laryngoscope. Anesthesia was maintained with sevoflurane (0.8–1.0%) in an oxygen-air mixture and continuous remifentanil infusion. Standard intraoperative monitoring included electrocardiography, noninvasive blood pressure, pulse oximetry, temperature, and bispectral index (BIS) monitoring. The BIS value was maintained between 40 and 60 throughout the procedure.

The intraoperative course was uneventful, with stable hemodynamic and respiratory parameters. The tracheostomy was completed in 30 minutes without anesthetic or surgical complications. At the conclusion of surgery, the patient was transferred to the intensive care unit under mechanical ventilation without reversal of neuromuscular blockade. After five days of postoperative intensive care, he was successfully weaned from mechanical ventilation. He was transferred to the general ward on postoperative day 7 and was discharged home on postoperative day 10 without perioperative seizure exacerbation or other complications.

DISCUSSION

Lennox–Gastaut syndrome (LGS) is a severe developmental epileptic encephalopathy characterized by multiple seizure types, cognitive impairment, and characteristic electroencephalographic abnormalities [3]. Although LGS is primarily managed medically, patients frequently require anesthesia for diagnostic or surgical procedures because of associated neurological and respiratory comorbidities. Perioperative management is particularly challenging because of refractory epilepsy, chronic use of multiple antiepileptic medications, developmental delay, and patient-specific airway or respiratory abnormalities. Consequently, anesthetic management should focus on maintaining seizure control while ensuring safe airway management and physiological stability.

One of the primary goals of anesthesia in patients with LGS is the prevention of perioperative seizure exacerbation. Seizures may be precipitated by hypoxia, hypercapnia, metabolic disturbances, inadequate anesthetic depth, surgical stress, or interruption of antiepileptic medications [4]. Continuation of regular antiepileptic medications throughout the perioperative period, including the scheduled morning dose on the day of surgery whenever possible, may reduce the risk of perioperative seizures. In the present case, pregabalin, valproate, and zonisamide were continued without interruption, and no seizures occurred during anesthesia or in the immediate postoperative period.

Long-term antiseizure therapy may also influence anesthetic management through pharmacologic interactions. Enzyme-inducing antiepileptic medications, such as carbamazepine and phenytoin, may increase the metabolism of anesthetic agents and reduce the duration of action of nondepolarizing neuromuscular blocking agents [5], whereas valproate has been associated with thrombocytopenia and coagulation abnormalities [6]. Although our patient was not receiving enzyme-inducing agents, recognition of these potential interactions remains important when planning anesthesia for patients with refractory epilepsy.

Selection of anesthetic agents should consider their influence on seizure threshold and their pharmacological interactions with antiepileptic medications. Propofol is widely regarded as an appropriate induction agent in patients with epilepsy because of its anticonvulsant effects and ability to suppress epileptiform activity [7]. Although seizure-like movements have occasionally been reported during induction or emergence, these events are uncommon, and propofol remains one of the preferred induction agents in epileptic patients. In our patient, propofol provided smooth induction without seizure-related complications.

Sevoflurane remains one of the most commonly used inhalational anesthetics in pediatric anesthesia because of its rapid onset and recovery and favorable airway profile. However, experimental and clinical studies have demonstrated that high concentrations of sevoflurane may induce epileptiform EEG activity, particularly in susceptible patients. Accordingly, maintenance with relatively low concentrations of sevoflurane combined with intravenous adjuncts has been recommended to minimize this risk while preserving the advantages of inhalational anesthesia [8]. In our case, anesthesia was maintained with 0.8–1.0% sevoflurane in combination with remifentanil, providing stable anesthetic depth without perioperative seizure activity.

Remifentanil was selected because its rapid onset, short context-sensitive half-life, and easy titration allow precise control of intraoperative analgesia while facilitating prompt postoperative neurological assessment. In addition, previous neuroanesthesia studies have shown that clinically relevant doses of remifentanil exert minimal effects on epileptiform cortical activity, making it an appropriate adjunct in patients with epilepsy [9]. Combined with low-dose sevoflurane, remifentanil allowed adequate analgesia and hemodynamic stability while reducing the requirement for higher concentrations of volatile anesthetic.

Airway management represented another important consideration in this patient. Although LGS itself is not intrinsically associated with difficult airway anatomy, generalized hypotonia, impaired upper airway tone, and associated craniofacial abnormalities may increase the likelihood of airway management difficulties in selected patients. Our patient demonstrated micrognathia, retrognathia, and dynamic upper airway obstruction on preoperative flexible fiberoptic laryngoscopy. These findings enabled careful preoperative planning, including preoxygenation with HFNC oxygen therapy, preparation of advanced airway devices, and implementation of a double-set-up strategy with immediate availability of emergency surgical airway access. Tracheal intubation was successfully accomplished on the first attempt using a video laryngoscope, suggesting that meticulous preparation can contribute to safe airway management even in patients with anticipated airway difficulty.

Overall, the favorable perioperative outcome in this case was likely attributable to multiple complementary strategies rather than any single intervention. Continuation of antiepileptic medications, appropriate selection and titration of anesthetic agents, vigilant physiological monitoring, and comprehensive airway preparation together enabled uneventful anesthetic management without perioperative neurological or respiratory complications.

CONCLUSION

General anesthesia can be safely administered in pediatric patients with Lennox–Gastaut syndrome through individualized perioperative planning. Continuation of antiepileptic medications, careful selection of anesthetic agents, and thorough evaluation and preparation for patient-specific airway abnormalities contributed to the successful outcome in this case. This report highlights the importance of comprehensive perioperative assessment and multidisciplinary planning in optimizing anesthetic safety for patients with LGS.

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