

Anesthesiologist's Concerns About Dandy-Walker Syndrome: Airway Management, Muscle Relaxants, and Train-of-Four Monitoring of Neuromuscular Blockade

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Abstract

Dandy-Walker syndrome is a rare congenital malformation of the posterior fossa that presents unique anesthetic challenges, including difficult airway management, altered consciousness, hydrocephalus, and potential for prolonged postoperative ventilation. Hydrocephalus, the most common finding, can complicate ventilation and intubation, whereas brainstem involvement, along with agenesis of the corpus callosum, pontine hypoplasia, and distortion of the medullary respiratory centers, may contribute to prolonged postoperative ventilation. Anesthetic management in such cases requires thorough airway assessment, preparation for potential airway difficulties, intracranial pressure monitoring and control, and total intravenous anesthesia (TIVA) to facilitate rapid emergence. We describe the case of a 3-year-old male with macrocephaly, movement disorders, delayed cognitive development, and altered mental status, scheduled for ventriculoperitoneal shunt placement under TIVA without muscle relaxants, with careful airway preparation and intracranial pressure control. To our knowledge, this is the first reported case of Dandy-Walker syndrome managed in this manner.

Keywords: Dandy-Walker syndrome; Pediatric neuroanesthesia; Intracranial pressure; Airway; Postoperative respiratory complications; Total intravenous anesthesia; Muscle relaxants; Train of four monitoring

Introduction

Dandy-Walker syndrome is a rare congenital malformation of the posterior fossa, characterized by hydrocephalus, dilatation

of the fourth ventricle, and hypoplasia of the cerebellar vermis. It is associated with increased intracranial pressure and neurological impairment. The estimated incidence is approximately 1 in 30,000 live births [1]. In addition to the characteristic central nervous system abnormalities, patients with Dandy-Walker syndrome may present with a range of associated congenital malformations, most commonly involving the cardiovascular and craniofacial systems. Several studies have documented the coexistence of non-cyanotic congenital heart diseases, such as atrial and ventricular septal defects, as well as more complex lesions, including tetralogy of Fallot. Craniofacial anomalies, along with musculoskeletal abnormalities such as scoliosis, have also been reported, reflecting the multisystem nature of the syndrome and underscoring the need for comprehensive multidisciplinary assessment [2-4].

Hydrocephalus represents the most common neurological manifestation, occurring in approximately 70-90% of cases. This frequently necessitates early neurosurgical intervention due to progressive intracranial hypertension and its adverse effects on neurodevelopment. Encephalocele is reported in about 16% of patients, while structural abnormalities of the corpus callosum, including hypoplasia or complete agenesis, are found in nearly 30%, further contributing to the complexity of the neurodevelopmental profile [4].

Clinically, Dandy-Walker syndrome is characterized by signs of increased intracranial pressure, macrocephaly, headache, nausea, and vomiting, alongside cerebellar signs such as movement disorders, respiratory difficulties, diminished gag reflex, poor nutritional status, and delayed neurodevelopment. Hydrocephalus remains the main surgical indication, with ventriculoperitoneal shunt placement being the standard approach.

We present the case of an infant diagnosed with Dandy-Walker syndrome within the first months of life after seizure onset. The diagnosis was confirmed through imaging and clinical evaluation. The patient underwent ventriculoperitoneal shunt placement under general anesthesia. In this case, particular emphasis was placed on the use of total intravenous anesthesia (TIVA) and the deliberate avoidance of muscle relaxants to address the potential for difficult endotracheal intubation, facilitate rapid emergence, and reduce the risk of postoperative pulmonary complications. This approach underscores the importance of tailoring anesthetic management to the unique neuroanatomical and respiratory challenges presented by Dandy-Walker syndrome.

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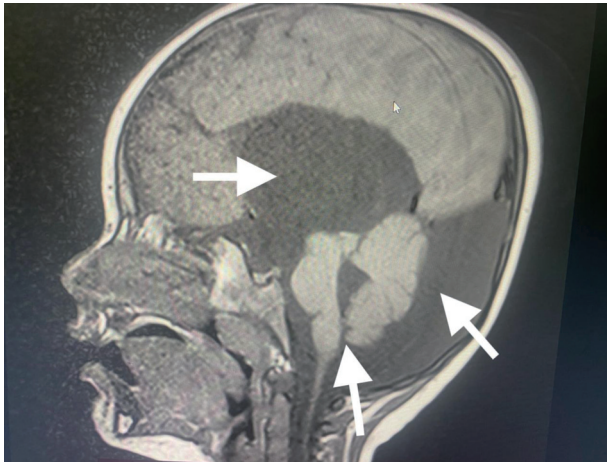


Figure 1. Preoperative head MRI demonstrating agenesis of the cerebellar vermis, dilatation of the fourth ventricle, and hydrocephalus (arrows). MRI: magnetic resonance imaging.

Case Report

Investigations

A 3-year-old male patient, weighing 21 kg, was admitted to our institution for surgical management of Dandy-Walker syndrome. The planned procedure was a ventriculoperitoneal shunt placement. The patient was born at term, with the first clinical signs of the condition appearing at 2.5 years of age, prompting referral to a pediatric neurologist and subsequent imaging studies. The diagnosis of Dandy-Walker syndrome was based on the presence of hydrocephalus, dilation of the fourth ventricle, and cerebellar hypoplasia (Figs. 1-3). Magnetic resonance imaging (MRI) concluded on agenesis of vermis, dilatation of the fourth ventricle, and hydrocephalus due to aqueductus Silvi stenosis.

The clinical presentation was dominated by signs of increased intracranial pressure, including agitation, altered mental status, nausea, vomiting, deviated eyes, and impaired

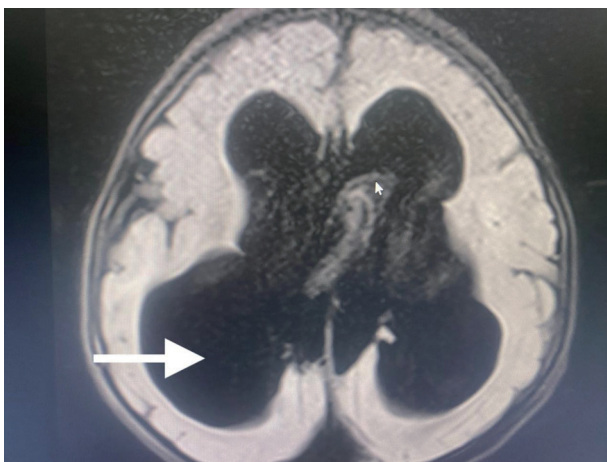


Figure 2. Preoperative head MRI demonstrating significant hydrocephaly (arrow). MRI: magnetic resonance imaging.

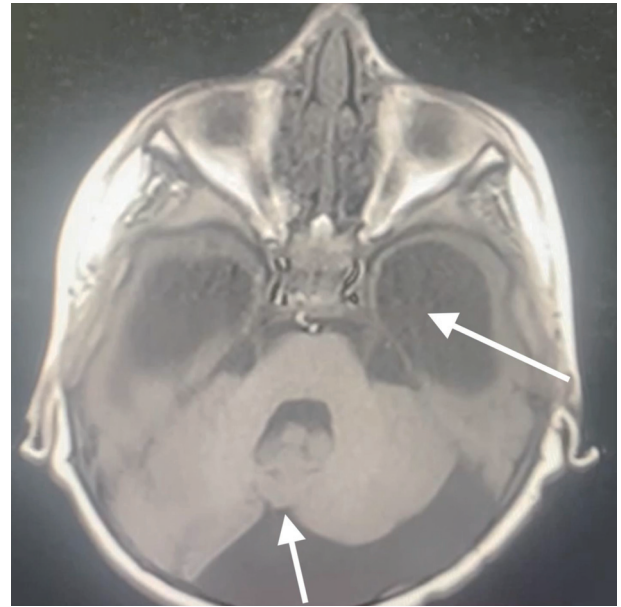


Figure 3. Preoperative head MRI demonstrating agenesis of the cerebellar vermis, significant hydrocephaly (arrows). MRI: magnetic resonance imaging.

motor coordination (Fig. 4). The patient could not follow simple commands, moderate ataxia, and manifested spastic tetra paresis especially in inferior limbs. Pediatric and cardiologic

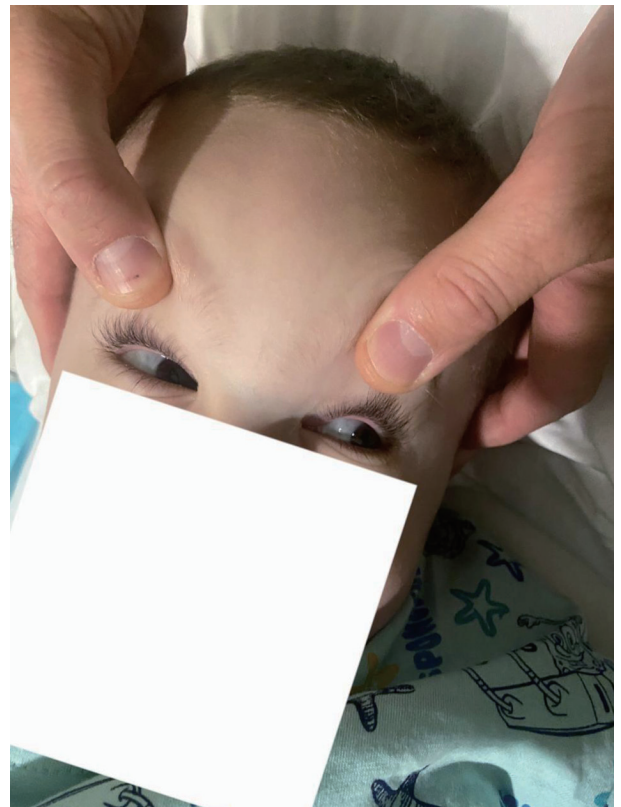


Figure 4. Eye deviation.



Figure 5. Mild micrognathia.

evaluations were unremarkable, and biochemical analyses were within normal limits. Airway assessment was limited by the patient's lack of cooperation; Mallampati class II-III and mild micrognathia was estimated (Figs. 5 and 6).

Diagnosis

No premedication was administered due to the patient's decreased level of consciousness and the associated risk of aspiration during anesthesia induction. Standard plus intracranial pressure monitoring was applied, and a 22-gauge peripheral intravenous cannula was inserted (Figs. 7 and 8). The initial attempt at tracheal intubation using conventional laryngoscopy with a Miller blade was unsuccessful. The airway was subsequently secured using a video laryngoscope (Karl-Storz, C-MAC 8043ZX) and trachea was intubated with size 5 mm internal diameter endotracheal tube (Fig. 9).

Treatment

General anesthesia was induced with intravenous administration of propofol at a dose of 2 mg/kg and fentanyl at 3 µg/kg to ensure adequate hypnosis and analgesia. Anesthesia maintenance was performed using TIVA, consisting of continuous infusions of propofol at 200 µg/kg/min and remifentanyl at 0.3 µg/kg/min, allowing for stable hemodynamic conditions and rapid emergence. No neuromuscular drugs were used. The surgical procedure was completed without intraoperative



Figure 6. Mallampati class II-III.

complications. Upon completion of surgery, the patient met extubation criteria and was successfully extubated in the operating room. No episodes of apnea or respiratory failure were observed during the 30-min observation period in the operating room following extubation.

Follow-up and outcomes

Postoperatively, the patient was transferred in stable condition to the ward for further monitoring and recovery. After postoperative imaging (head MRI) and abdominal ultrasound was performed, the patient was discharged home (Figs. 10 and 11). Informed consent was obtained from the patient's relative, and all images are provided courtesy of the American Hospital imaging database.

Discussion

Dandy-Walker syndrome represents a complex spectrum of intracranial malformations with significant clinical and anesthetic implications. The constellation of anomalies, including hydrocephalus, cerebellar vermis hypoplasia, and cystic dilation of the fourth ventricle, presents multiple perioperative challenges. Anesthetic management requires preparedness for elevated intracranial pressure, difficult airway scenarios often compounded by craniofacial dysmorphisms, and potential postoperative respiratory complications related to brainstem dysfunction. Pediatric patients further require specialized expertise, particularly in



Figure 7. ICP monitoring (ICP was 15 mm Hg, in arrow). ICP: increased intracranial pressure.



Figure 8. Standard monitoring.



Figure 9. Video laryngoscopy intubation.

establishing and maintaining peripheral venous access. Comprehensive preoperative assessment and multidisciplinary planning are essential to optimize outcomes [5, 6].

While principles of pediatric neurosurgical anesthesia resemble those in adults, unique challenges arise due to congenital anomalies, distinct tumor types, and variable anatomical localizations. Key considerations include monitoring and controlling intracranial pressure, managing difficult airways, securing venous access, maintaining age-appropriate mean arterial pressure, and preventing hypoxia or hypercapnia [6]. Our case involved a 3-year-old male born at term via elective cesarean section, who presented with seizures shortly after birth and was treated with valproate and levetiracetam. Neurodevelopment was nearly normal until age 2, after which progressive

macrocephaly, developmental delay, recurrent pneumonia, and impaired motor function became evident as a pattern consistent with Dandy-Walker syndrome. A thorough pediatric cardiac evaluation, including examination, electrocardiography, and echocardiography, revealed no abnormalities [7].

Airway evaluation is critical in patients with craniofacial or neurodevelopmental abnormalities. Features such as macrocephaly, micrognathia, macroglossia, and cervical spine anomalies can increase the risk of difficult mask ventilation and intubation [8, 9]. In our patient, mild micrognathia and a Mallampati class II-III score were noted. Due to develop-

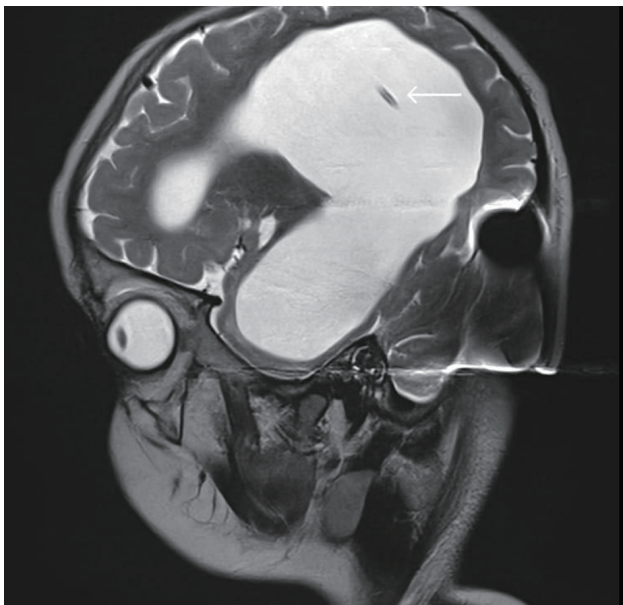


Figure 10. Postoperative MRI indicating shunt catheter in arrow. MRI: magnetic resonance imaging.

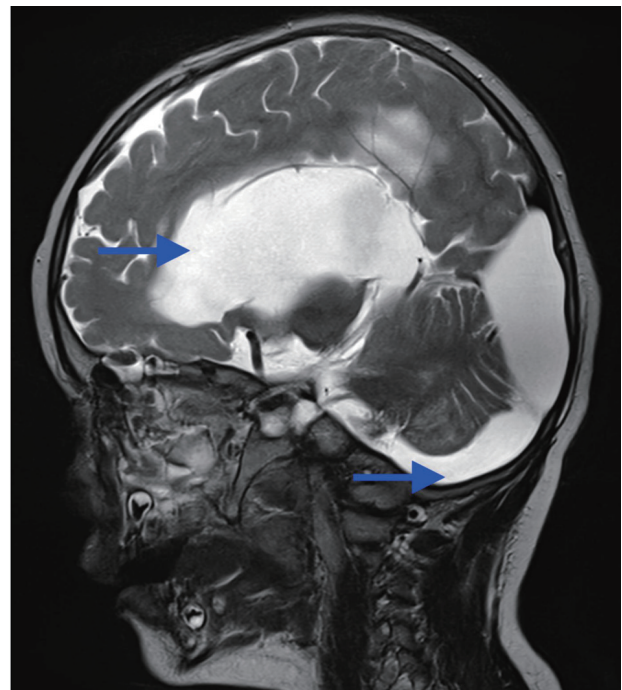


Figure 11. Postoperative MRI indicating reduced hydrocephaly in arrows. MRI: magnetic resonance imaging.

mental delay and limited cooperation, comprehensive airway assessment was incomplete [9]. After induction, conventional Miller blade intubation failed, but successful endotracheal intubation was achieved with a pediatric video laryngoscope, facilitating atraumatic airway management.

Postoperative respiratory failure remains a major concern, as central nervous system (CNS) abnormalities including corpus callosum agenesis, pontine hypoplasia, and medullary respiratory center distortion may impair respiratory drive and airway reflexes [10]. Laryngospasm, which can be more frequent and severe in patients with brainstem lesions, did not occur in our patient [10]. To mitigate these risks, we used TIVA with short-acting agents, avoiding neuromuscular blocking drugs when train-of-four monitoring was unavailable. This strategy allowed rapid emergence, safe extubation, and preservation of spontaneous respiration.

Reversal of neuromuscular blockade is a critical step in anesthesia to ensure safe postoperative recovery when muscle relaxants are used. Two primary approaches, quantitative train-of-four monitoring and clinical judgment, are used to assess residual blockade. TOF monitoring provides objective, real-time data that improve the accuracy of reversal agent dosing and reduce the risk of postoperative respiratory complications [11]. Clinical judgment relies on observable patient responses and is immediately available but is subjective and less sensitive in detecting residual paralysis. Another meta-analysis of 24 trials (3,375 patients) evaluated the impact of intraoperative neuromuscular monitoring (NMM) on postoperative residual curarization (PORC). PORC incidence was lower with intermediate-acting neuromuscular blockers compared to long-acting agents. Monitoring, whether via simple peripheral nerve stimulator or objective device, did not significantly reduce PORC. Mivacurium was excluded due to limited data [12]. PORC remains common, and the effectiveness of intraoperative NMM has been unclear. This meta-analysis of 53 studies (12,664 patients) found that quantitative NMM significantly reduced PORC incidence compared to qualitative or no monitoring, particularly with intermediate-duration neuromuscular blocking agents [13]. Pediatric patients are at risk of PORC due to age-related differences in the pharmacokinetics and pharmacodynamics of neuromuscular blocking agents (NMBAs) and the short duration of many surgeries [14]. This meta-analysis of 71 trials including 4,319 participants quantified spontaneous recovery times (t₂₅, RI₂₅₋₇₅, tTOF₉₀) across different NMBAs and age groups. Recovery was faster in older children compared to neonates and infants, with duration further prolonged by volatile anesthetics and aminosteroid NMBAs. The findings highlight significant variability in neuromuscular recovery, underscoring the need for quantitative NMM even after a single NMBA dose in pediatric patients [14]. The patient remained stable in the postanesthesia care unit (PACU) for 30 min with no signs of respiratory depression.

Based on this experience, ultrashort-acting agents such as propofol and remifentanyl, combined with avoidance of muscle relaxants, are recommended to minimize postoperative respiratory complications [15]. Interesting findings were reported by Lee et al. In their retrospective study, volatile anesthetics showed no superiority over TIVA in reducing postoperative pulmonary complications (PPCs) after anatomical pulmonary

resection in patients with lung cancer [16]. Recent evidence supports the superiority of total TIVA. In a randomized controlled trial, Chang et al compared TIVA with inhalational anesthesia, using postoperative pulmonary complications (PPCs) as the primary endpoint. Their results demonstrated that TIVA significantly reduced the incidence of PPCs [17]. Given the respective advantages of each anesthetic modality, the choice of anesthesia should be tailored to individual patient risk factors and clinical circumstances [17]. Kannabiran et al, in their narrative review, highlighted TIVA as a favorable option due to its smooth induction, faster recovery, reduced postoperative nausea and vomiting, and its capacity to optimize surgical conditions [18]. Kumaria et al similarly reported superior perioperative outcomes with TIVA, attributing the benefit largely to better intraoperative brain relaxation [19]. Oh et al published their data of a retrospective cohort study of 144,506 adults undergoing cranial neurosurgery in South Korea (2016 - 2021) compared TIVA with inhalation anesthesia. The 90-day mortality rates were similar between groups (14.0% TIVA vs. 14.2% inhalation; odds ratio (OR) 0.97, 95% confidence interval (CI) 0.94 - 1.01, P = 0.188). However, TIVA was associated with a significantly lower rate of postoperative complications (47.1% vs. 50.3%; OR 0.88, 95% CI 0.86 - 0.90, P < 0.001). Thus, while anesthetic type did not affect short-term mortality, TIVA was linked to reduced postoperative morbidity [20].

Effective postoperative pain management is essential. Opioid use may be limited due to potential respiratory depression and altered consciousness, while NSAIDs are constrained by bleeding risk. Multimodal analgesia, including regional techniques, NSAIDs, dexmedetomidine, paracetamol, ketamine, and scalp blocks, is effective for controlling postoperative pain, reserving opioids for insufficient pain control [21-24].

Generally, careful evaluation by both neurosurgeons and anesthesiologists is essential to ensure patient safety and optimize prognosis. This assessment begins with determining the indication and timing of surgery, which should be guided by an individualized approach from both surgical and anesthetic teams. In children with posterior fossa arachnoid cysts, management decisions should be based primarily on clinical presentation rather than radiological findings. Pesaresi et al found that a “wait and see” approach is effective for asymptomatic patients, while surgical decisions should consider cyst location, hydrocephalus, and patient age. Age < 12 months, hydrocephalus, macrocephaly, and shunt placement increase the risk of re-surgery. Outcomes were similar across microsurgical, endoscopic, and shunting procedures, highlighting the importance of individualized treatment planning [25].

Our anesthetic strategy, summarized in Table 1, emphasized thorough preoperative evaluation, exclusion of additional congenital anomalies, careful airway management, and prevention of postoperative respiratory complications. Difficult airway protocols were established, and postoperative respiratory care included avoidance of muscle relaxants or the use of train-of-four NMM when relaxants were necessary.

Conclusions

Pediatric neuroanesthesia requires dual expertise in both

Table 1. Key Considerations and Recommended Actions in Pediatric Neuroanesthesia for Patients With Dandy-Walker Syndrome [7-9]

Situation	Management considerations	Recommended actions
Pediatric neuroanesthesia expertise	Requires specialized pediatric and neuroanesthetic skills	Fellowship or extended training programs for advanced experience
Airway management	1) Macrocephaly, micrognathia, macroglossia; 2) Cervical spine anomalies; 3) Limited cooperation in pediatric patients.	1) Thorough preoperative airway assessment; 2) Difficult airway protocol in place; 3) Consider laryngeal mask airway or video laryngoscope if conventional intubation fails.
Intubation	High risk of failed conventional intubation	1) Prepare alternative devices; 2) Ensure rapid, atraumatic intubation
Anesthetic technique	1) Risk of postoperative respiratory complications; 2) Brainstem dysfunction.	1) Total intravenous anesthesia with short-acting agents; 2) Avoid muscle relaxants if train-of-four monitoring unavailable; 3) Ensure rapid emergence.
Postoperative respiratory care	Risk of apnea, aspiration, atelectasis, prolonged ventilation	1) Close monitoring in postanesthesia care unit; 2) Respiratory exercises as indicated; 3) Train-of-four monitoring if muscle relaxants used.
Pain management	Limitations due to respiratory depression and bleeding risk	1) Multimodal analgesia (nonsteroidal anti-inflammatory drugs, paracetamol, ketamine, dexmedetomidine, regional techniques); 2) Reserve opioids for insufficient pain control.
Cardiac and other congenital anomalies	Possible coexisting defects	Comprehensive preoperative evaluation including electrocardiography and echocardiography

pediatric and neuroanesthetic management. Patients with Dandy-Walker syndrome present unique perioperative challenges, necessitating comprehensive preoperative evaluation. Special attention should be paid to associated congenital anomalies, particularly cardiac malformations, airway abnormalities, and neurologically related dysfunctions. Careful airway management, including a clearly defined rescue strategy, is essential, along with the use of short-acting anesthetic agents to allow precise control of anesthesia. Muscle relaxants should be avoided if train-of-four monitoring is unavailable, and rapid emergence from anesthesia should be planned to minimize postoperative respiratory complications. The use of total intravenous anesthesia under these principles is recommended to ensure a safe and uneventful anesthetic course.

Learning points

Pediatric neuroanesthesia is a highly specialized field that demands a skilled and experienced multidisciplinary team.

Dandy-Walker syndrome poses distinct challenges, particularly concerning airway management, intracranial pressure control, and the prevention of postoperative respiratory complications.

The anesthesia plan prioritized difficult airway management, minimized pulmonary complications with short-acting intravenous agents, avoided muscle relaxants, when possible, used train-of-four monitoring if relaxants were necessary, and ensured rapid emergence.

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Financial Disclosure

None to declare.

Conflict of Interest

None to declare.

Informed Consent

Informed consent has been obtained.

Author Contributions

RD, AA, and GH: manuscript writing; OD and FC: literature searching; RD, AD, and KL: language editing.

Data Availability

The data supporting the findings of this study are available

from the corresponding author upon reasonable request.

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