



Sugammadex and General Anesthesia in Two Pediatric Patients with Joubert Syndrome: Case Report

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CASE REPORT

ABSTRACT

Joubert syndrome (JS) is a rare genetic disorder that affects the cerebellum, controlling balance, and coordination, and it usually presents a molar tooth sign on magnetic resonance imaging. It is characterized by ataxia, an abnormal breathing pattern referred to as hyperpnea, sleep apnea, hypotonia, and abnormal eye and tongue movements. Because of sensitivity to respiratory depressant agents, including opiates, nitrous oxide, and neuromuscular blockers, and potential difficult airways, anesthetic management requires attention. Sugammadex binds to rocuronium and is a reverse neuromuscular blocker. In this study, two cases of JS where sugammadex was used for general anesthesia are presented. Sugammadex may be a safe general anesthetic for patients with JS.

Keywords: General anesthesia, sugammadex, Joubert syndrome

INTRODUCTION

Joubert syndrome (JS), first described in 1968, is a rare autosomal recessive disorder that affects the cerebellum and brain stem, which are important organs for an anesthesiologist (1). The syndrome is characterized by hypotonia, breathing abnormalities, oculomotor apraxia or other abnormal eye movements, severe psychomotor retardation, ataxia, facial dysmorphism, lobulated tongue, cleft lip and palate, and scoliosis. Cerebellar abnormalities together with vermian agenesis represent the clinical aspect of respiratory impairment, particularly in the neonatal period (2). Because of patient anatomic features, including palatal malformations, laryngomalacia, and micrognathia, airway management may be difficult and respiratory abnormalities may cause problems before and during anesthesia (3). Patients with JS are sensitive to the respiratory depressant effects of anesthetic agents, including nitrous oxide (N₂O), neuromuscular blockers, and opioids (4, 5). Sugammadex is a novel agent for the reversal of the neuromuscular blockade that provided by rocuronium and vecuronium. It acts differently from anticholinesterases and can reverse even profound blocks induced with vecuronium or rocuronium. The neuromuscular block can be reversed at any time by the administration of cysteine, which could significantly reduce the risk of postoperative residual neuromuscular blockade (6). Sugammadex binds to rocuronium molecules in a 1:1 ratio (7). This feature makes sugammadex preferable for the management of difficult airways as it rapidly eliminates neuromuscular blockades (8).

In this case study, we report the successful application of sugammadex for dental procedures under general anesthesia in two pediatric patients with JS.

CASE REPORTS

Case 1

An 11-year-old boy (weight: 40 kg) presented to our clinic for a dental procedure requiring general anesthesia. Previously, he had undergone other surgeries due to bilateral inguinal hernia and cataract at 4 and 10 years of age. As learned from his history, he had received general anesthesia for these surgeries without any complication in the postoperative period. On conducting a physical examination, there was corpus callosum agenesis, dental anomalies, mental-motor retardation, lower extremity paraplegia, mild scoliosis, and bifid uvula. The Mallampati score was class II. White blood count, platelet count, and electrolyte concentrations were within normal limits.

Case 2

A 9-year-old boy (weight: 50 kg) presented to our clinic for a dental procedure requiring general anesthesia. Previously, he had received sedation for an eye examination and had undergone hearing tests without any complication. He was the third child in his family and had two older sisters with JS. He had mental-motor retardation, intellectual disability, hypotonia, abnormal eye movements, developmental delay as a newborn, moderate hearing loss, autism, and a "molar tooth sign" on performing brain imaging. Blood evaluations and electrolyte concentrations were normal. He had no facial abnormalities, and his Mallampati score was class II.

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Anesthesia technique

An informed consent was taken from the parents before the procedure. The patients were taken into the operating room after intravenously midazolam premedication, and routine monitors were applied. Following preoxygenation, Pofol (propofol, 3 mg/kg) and Esmeron (rocuronium bromide, 0.5 mg/kg) were injected for neuromuscular blockade to facilitate intubation. Sevoflurane was used for the induction and maintenance of anesthesia at a dose of 1.5 minimum alveolar concentration (MAC). The epiglottis could be easily seen during direct laryngoscopy and was graded as I in both patients. No opiate was applied during induction and for the maintenance of anesthesia. Direct laryngoscopy was performed using a Macintosh blade, and the trachea was orally intubated with 6.5-mm and 5.5-mm tubes for cases 1 and 2, respectively. Rightward cricoid pressure was needed to bring the posterior portion of vocal cords into the visualization field. No other anesthesia-related difficulties were noted. Sevoflurane (1.5 MAC) inhalation in oxygen:air mixture was used during the surgery. Analgesia was achieved with installation of 2-4 mL of articaïne (Ultracain DS) to each tooth. The dental procedures lasted for 90 and 65 min in cases 1 and 2, respectively. At the end of the surgery, 4 mg/kg of sugammadex (Bridion) was used as an antidote for rocuronium bromide. Sugammadex rapidly reversed the block, and endotracheal tubes were removed when the patients began to breathe regularly and adequately. After the extubation, hemodynamic and respiratory functions were stable in both patients. No complications were seen after the extubation. The patients were transferred to the ward while breathing spontaneously and peripheral oxygen saturation as being 98% in air room without taking supplemental oxygen, and were monitored with a pulse oximeter. There were no apneic or hyperpneic periods during the postoperative follow-up. Oral intake was began with liquids 3 hours after the procedure. Both patients were discharged home without any respiratory or other complications.

DISCUSSION

Joubert syndrome is a rare genetic disorder that affects the area of the brain that controls balance and coordination (in particular, the cerebellum) and presents a “molar tooth sign” on magnetic resonance imaging. It is characterized by ataxia (lack of muscle control), an abnormal breathing pattern referred to as hyperpnea, sleep apnea, hypotonia, a characteristic facial appearance, microcephaly, and abnormal eye and tongue movements. Mild or moderate retardation, extra fingers and toes, cleft lip and palate, irregular tongue protrusion, and seizures may also be present in many patients (9).

The first report about anesthetic management in JS was published in 1989. The authors described opiate sensitivity and abnormal control of respiration (10). In many studies, it has been reported that opiates must be avoided because patients with JS have abnormalities in respiratory control resulting in episodic tachypnea and apnea and a susceptibility to the respiratory depressant effects of anesthetic agents (3, 4). In our patients, there was no respiratory abnormality or difficult airway during the preoperative period. Even so, we did not use opioids during the induction and maintenance of anesthesia due to potential adverse effects on respiratory patterns related to JS.

In 2006, Platis et al. (11) reported a 19-year-old woman who underwent general anesthesia with propofol, fentanyl, and atracurium for peritoneal dialysis catheter insertion. The authors observed apneic episodes after the extubation. While abnormal breathing wanes in adults, they suggested being aware of postoperative complications in patients with JS and avoiding opioids (11). In that study, the authors confirmed muscle relaxation using a nerve stimulator and extubated the patient using clinical criteria. In our patients, general anesthesia was induced with propofol and was maintained with sevoflurane without N₂O in oxygen. Rocuronium bromide (0.5 mg/kg) was used to facilitate tracheal

intubation. In contrast, we used sugammadex (4 mg/kg) intravenously for reversing muscle relaxation. Abrishami et al. (12) suggested that in comparison to a placebo or neostigmine, sugammadex can more rapidly reverse rocuronium-induced neuromuscular blocks regardless of the depth of the block. They found that sugammadex was associated with serious adverse events in less than 1% of the patients. We used sugammadex because neuromuscular blockers may cause residual blocks after general anesthesia is completed and this may aggravate the respiratory abnormalities in patients with JS. Postoperatively, no respiratory complications, including hyperpneic or apneic episodes, were observed.

Patients with JS are susceptible to respiratory depressant agents such as opioids, N₂O, and neuromuscular blockers. Thus, anesthetic management in JS needs more attention related to ventilation parameters and potential difficult airways. In conclusion, we found that general anesthesia with sevoflurane, rocuronium bromide, and sugammadex may be a safe and reliable method for patients with JS. Further studies are needed to clarify the use of sugammadex in JS for general anesthesia.

Informed consent: Written informed consents were taken from the parents.

Authors' Contributions: Conceived and designed the experiments or case: DGC, SD, HP, MC. Performed the experiments or case: DGC, MDY, SD. Analyzed the data: DGC, FD, SD. Wrote the paper: DGC, FD. All authors have read and approved the final manuscript.

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