





## CASE REPORT

# Postoperative Bronchospasm in a Child with Coffin-Siris Syndrome Following Administration of Sugammadex

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## Abstract

A 13-year-old boy with Coffin-Siris syndrome was scheduled to undergo dental surgery for dental treatment with general anesthesia. Mental retardation, finger anomalies and coarse facies features were recorded in preoperative assessment. Propofol, Fentanyl and Rocuronium were given for anesthesia induction. Sevoflurane was used as an inhalation anesthetic drug in anesthesia maintenance. At the end of the operation, the patient was extubated using sugammadex as a neuromuscular reversal drug. After the extubation, bronchospasm appeared. The patient treated for bronchospasm. At the postoperative care unit, the patient's vital signs were followed for a while and discharged on postoperative day without any problems. For the reason those pulmonary problems of the patients with Coffin-Siris syndrome which was unknown or underestimated before anesthesia, sugammadex administration is a risk for bronchospasm. Consequently, pre-anesthetic evaluation should be performed carefully in patients with Coffin-Siris syndrome to be treated with sugammadex and notably respiratory pathologies should be questioned.

## Keywords

Postoperative bronchospasm, Coffin-Siris syndrome, Sugammadex, Dental surgery

## Key Message

Sugammadex is a risk factor for bronchospasm in patients with Coffin-Siris syndrome.

growth restriction, absent fifth-digit fingernails or hypoplastic fifth finger terminal phalanx, coarse facial features, hirsutism/hypertrichosis, sparse scalp hair, cardiac, neurological, gastrointestinal, genitourinary anomalies [1]. Since poor communication, dental interventions are difficult in these patients. Whereas general anesthesia is preferred in such patients without sufficient mouth opening, this technic carries many risks such as difficult intubation and bronchospasm due to the underlying anomalies in these patients.

Use of sugammadex, as a neuromuscular reversal drug during general anesthesia is increasingly preferred due to lack of direct interaction with the cholinergic system [2]. However, recent studies on sugammadex have reported that it may cause upper airway obstruction, such as laryngospasm [3] or bronchospasm [4]. For this reason, the pulmonary problems are challenging for the anesthesiologist.

It is well recognized that patients with preexisting pulmonary disease have an increased risk of perioperative pulmonary complications [5,6]. Hence we aimed to report bronchospasm after sugammadex without significant respiratory pathology in a patient with CSS, not yet reported in the literature.

## Introduction

Coffin-Siris syndrome is a rare genetic disorder that causes mental and developmental retardation such as

## Case Presentation

A 13-year-old boy with Coffin-Siris syndrome, a height of 105 cm and weighs 35 kg was scheduled to undergo dental surgery for dental restoration with gen-

eral anesthesia at an ambulatory setting and same-day admission. In preoperative assessment, mental retardation, cognitive disability, hypoplastic fifth-finger terminal phalanx, coarse facies reflected by flat nasal bridge, wide mouth, thick lips, macroglossia, micrognathia, and irregular teeth were recorded. Airway evaluation was not performed due to the lack of cooperation of the patient. Pulse oximetry, noninvasive blood pressure, and electrocardiography monitoring were performed continuously. After preoxygenation during 5 minutes, anesthesia was induced with 100 mg Propofol intravenous (IV), 50 mcg Fentanyl IV and after successful bag-mask ventilation via face mask, rocuronium 25 mg IV was given. The laryngeal view during direct laryngoscopy was Cormack-Lehane score 2. Cuffed endotracheal tube number 6 mm internal diameter was safely placed into the trachea without trouble. Afterward, endotracheal intubation was confirmed by chest movement, auscultation, and end-tidal CO<sub>2</sub> waveform. Anesthesia was maintained with Sevoflurane 50% oxygen/air. During operation vital signs were stable. One root canal treatment, 3 teeth extractions, 4 teeth fillings were done during general anesthesia. Paracetamol 150 mg IV was given for postoperative analgesia. At the end of the operation sugammadex, 100 mg IV was given for neuromuscular block reversing. Since the patient regained clear consciousness and spontaneous breathing at 10 mL/kg, the intubation tube was removed without any problem. He had a small amount of secretion so oral aspiration was done gently. About three minutes later after extubation, respiratory distress began. The patient had tachypnea and dyspnea. Afterward, oxygen saturation decreased quickly to the critical blood oxygen saturation level of 5% (SpO<sub>2</sub>). Immediately 100% oxygen at 10 ml/min bag-valve-mask ventilation was started. The lung sounds presented expiratory wheeze, prolonged exhalation on auscultation. The patient therefore presumptively treated for bronchospasm by deepening the plane of anesthesia with Propofol and treating with inhaled beta-2 agonists and intravenous steroids. Eventually, hypoxemia was resolved slowly. At the postoperative care unit, the patient's vital signs were followed for a while and discharged on postoperative day without any problems.

## Discussion

CSS is a rare syndrome and has reported in only a few patients since 1970. Because of some typical features such as craniofacial and systemic anomalies, patients with CSS can be a concern to anesthesiologists especially for general anesthesia. Above all pulmonary problems are important for such syndromic patients in the perioperative period for morbidity and mortality.

Increased airway sensitivity, which can be associated with upper respiratory tract infection, probably increases the risk of perioperative respiratory adverse events [7]. Most of the perioperative respiratory adverse

events can be anticipated, recognized, and treated easily, but rare ones such as bronchospasm that can lead to oxygen desaturation and death is a serious complication. Its prevention and treatment is challenging and a lifesaver at the same time for anesthesiologists.

Upper and lower respiratory tract infections are often seen in these patients [1]. Amao, et al. reported that sugammadex causes bronchospasm in patients with a history of respiratory disease [2]. In our patient, there were no signs and symptoms of respiratory tract infections on the day of surgery and recent days. When in fact, we learned from the family that the patient had had frequent upper respiratory infections in the last six months. According to the results of this case, the effects of airway infections may persist for a long time without any symptoms in patients with Coffin-Siris syndrome.

N Yashioka, et al. reported that the responsiveness of the bronchial smooth muscles to the acetylcholine was not affected by the *in vitro* sugammadex administration in rats [8]. On the other hand, some side effects had reported in studies of healthy volunteers and patients. Amao, et al. reported that patients with bronchospasm have a history of asthma [4]. In these patients with asthma history, sugammadex may cause bronchospasm in bronchial smooth muscles due to hypersensitivity. When in fact, delayed bronchospasm seen in the same study may be caused by sugammadex-rocuronium clathrate complex or mast cell stimulation. In our case, bronchospasm developed in a short time after the administration of sugammadex. It is a short time for the stimulation of the smooth muscle by the clathrate complex or activation of the mast cells. Here we think that smooth muscles are activated directly by sugammadex.

Successful anesthesia management has been published in a few reported cases in the literature in CSS and also the use of sugammadex has been reported in a few patients in CSS. No description of the complications in patients with Coffin-Siris syndrome associated with the sugammadex is in practice. Under those circumstances, this is the first case in Coffin-Siris syndrome to report bronchospasm after sugammadex.

## Conclusion

Reversing rocuronium-induced neuromuscular blockade with sugammadex is a risk factor for patients with Coffin-Siris syndrome having common pulmonary pathologies. For this reason, sugammadex should be used with proper caution in patients with underlying possible pulmonary pathologies.

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