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CASE REPORT

Management of Two Years Old Child with Russell-Silver Syndrome - A Case Report

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Abstract

A two-year-old boy with a known history of Russell-Silver syndrome was scheduled for elective surgery for hip spica. The patient had a history of mild developmental delay, failure to gain weight, and frequent vomiting. Anesthetic implications of RSS, including airway management, glucose homeostasis, hypothermia, and pharmacological considerations, were considered during the patient's pre-operative assessment and intraoperative management. The patient was induced with propofol and fentanyl, and maintenance anesthesia was achieved with sevoflurane. The patient's airway was safely secured, and extubation was performed smoothly. The anesthetic management proceeded without complications, ensuring a successful outcome. This case report highlights the importance of considering the specific anesthetic implications of RSS in the perioperative management of patients with this rare genetic syndrome.

Keywords: Child, Russell-Silver syndrome, Anesthesia, Micrognathism, Intubation.

Introduction

Russell-Silver syndrome (RSS) is a rare genetic malformation characterized by face and limb asymmetry with growth retardation.[1] It was first defined independently by Silver (1953) and Russell (1954) . The disease has a wide heterogeneous genetic spectrum. About 60% of patients have abnormalities involving chromosomes 7 or 11. Yet, in approximately 40% of patients with a definite clinical diagnosis of RSS, there is no known cause.[1] Affected individuals have a wide range of features affecting different body parts. The most common are craniofacial features, including a large-head-for-body, triangular-shaped face, relative macrocephaly, prominent forehead, and micrognathia.[1] This report focuses on a patient with RSS who underwent general anesthesia.

Consent for publication

Written informed consent was taken from the patient's father for this publication.

Case presentation

A two-year-old, 8kg male child with Russell-Silver syndrome was electively scheduled for hip spica

under general anesthesia. The patient suffers from mild developmental delay, and, according to the pediatrician, he only recently started to walk unaided and speak monosyllabically. The patient was born prematurely, admitted to the neonatal intensive care unit (NICU), and mechanically ventilated for two months. Until reaching the age of six months, he was kept on a nasogastric tube (NGT) due to failure to gain weight and frequent vomiting. Moreover, he was admitted to the hospital multiple times due to bronchopneumonia. As gastroesophageal reflex was suspected, he underwent a modified barium swallow, which returned negative for reflexes. A review of the systems showed no significant findings, except for the issues mentioned above. Moreover, the past surgical history of the patient was insignificant.

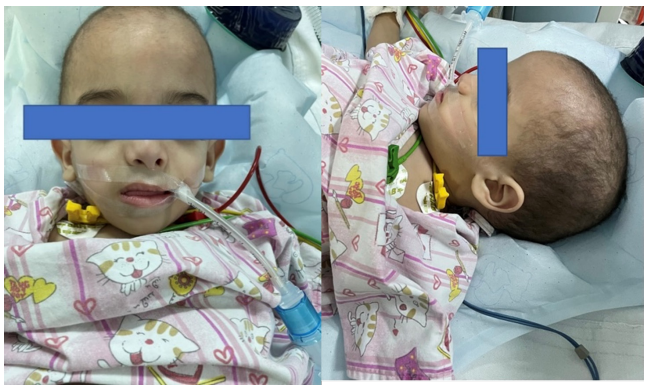

Figure 1: Frontal and lateral picture of the child with Russell-Silver

The patient presented with a typical triangular-shaped face with micrognathia during the pre-operative assessment. Vital signs revealed a heart rate of 112 beats/minute, blood pressure of 93/61, and temperature of 36.3 degrees Celsius. The patient received 5% dextrose with 0.5% normal saline and underwent frequent blood sugar monitoring; the last reading was 201mg/dl. The patient weighed 8 kg and measured 84 cm in height, resulting in a BMI of approximately 11.4, classifying them as underweight. Physical examination revealed unremarkable findings on cardiopulmonary examination, but mild hypotonia was present in the lower limbs. An airway exam confirmed a midline trachea, a Mallam-Pati classification of two, normal dentition, and adequate mouth opening. Due to the anticipated difficulty with intubation, a pediatric review was conducted to rule out any associated anomalies.

The patient underwent elective hip spica surgery with a 2.5mg oral midazolam pre-medication administered an hour before the procedure. Before surgery, the patient was cannulated with a 22 G cannula, and anesthesia was induced intravenously using 2mcg fentanyl and 20mg propofol without using muscle relaxants. During the 40-minute surgical procedure, anesthesia was maintained with a mixture of 2% sevoflurane, 30% oxygen, and the remaining air with a targeted minimum alveolar concentration (MAC) of no lower than 1. Adjuvant medications such as 2mg Dexamethasone and 80mg IV Perfalgan were subsequently administered.

Bag-mask ventilation during induction was smooth, and the patient was intubated with a size four tube using direct laryngoscopy with a fiberoptic scope on standby. A Bair Hugger and dextrose solution infusion was employed to prevent hypothermia and hypoglycemia.

The patient's airway was safely secured, and extubation was performed smoothly before transferring to the recovery room. The patient was closely monitored for 35 minutes in the recovery room before being transferred to the ward. During the stay, the patient received 4L/min oxygen via a face mask to maintain a saturation level of 96%, gradually decreasing to room air with a saturation level of 98%. Blood sugar and temperature levels were closely monitored throughout. Once the patient had fully recovered and awakened, he was transferred to the ward for further observation and care.

The anesthetic management proceeded without complications, ensuring a successful outcome.

Discussion

This two-year-old male child with Russell-Silver syndrome may face several problems during anesthesia before elective hip spica surgery. The patient's history of failure to gain weight and frequent vomiting may indicate underlying gastrointestinal issues that can affect the patient's ability to tolerate anesthesia. The patient's history of bronchopneumonia may also indicate possible respiratory issues. The patient's typical triangular-shaped face with micrognathia can make airway management and intubation difficult.[2] The patient's

mild hypotonia in the lower limbs may also affect muscle relaxation during anesthesia induction, which may result in profound relaxation due to small muscular mass.[3,5] Additionally, the patient's low body weight may increase the risk of hypothermia during surgery.[3] The patient's blood sugar levels also need to be closely monitored due to the risk of hypoglycemia, which is common in RSS patients.[3] These factors must be considered during the pre-operative assessment and anesthesia management to ensure a safe and successful surgery.

Russell-Silver Syndrome (RSS) is usually associated with various metabolic and phenotypic abnormalities,[2] of which are important to consider during pre-anesthetic preparation. In addition to the above-mentioned craniofacial features, RSS patients may suffer from gastrointestinal problems such as gastroesophageal reflux, poor feeding, and weight loss.[1] Thus, those patients are vulnerable to malnutrition, potentially impeding proper recovery post-surgery.[6] Moreover, Neurological problems can include hypotonia and delayed motor development.[1] There may also be cardiac defects, such as ventricular and atrial septal defects, and other issues, including limb abnormalities, genitourinary abnormalities, and various endocrinopathies.[1]

A major concern for these patients is the potential difficulty maintaining the airway due to facial dysmorphism (micrognathia, retrognathism, and hypognathous).[2,4] Mask ventilation can be affected by the small face/mouth.[2] Direct laryngoscopy may be difficult owing to a small mouth opening and small anterior larynx.[2] Tracheal intubation can be complicated by subglottic stenosis.[4] It is therefore recommended to prepare for potentially difficult airway management according to the algorithm. In addition, according to the child's age, it is suggested to prepare a smaller fiberoptic bronchoscope in case of difficult intubation.[2]

In our case, we anticipated difficulty in intubation from the pre-operative assessment. Therefore, various sizes of trachea tubes, smaller fiberoptic bronchoscopes, and a video laryngoscope were present. However, we didn't face difficulty during intubation, owing it also to the experience of the senior anesthetist.

In addition, special attention should be paid to abnormal glucose hemostasis, as hypoglycemia is the most common endocrinopathy in this group.[1] Two hypotheses for this have been suggested. First, hypoglycemia, particularly in those small-for-gestational-age neonates, may be caused by the fast depletion of limited hepatic glycogen stores.[3] Secondly, the disproportion between the sizeable cranial-to-body mass ratio increases glucose consumption.[3] Other possible factors could include prolonged fasting and peri-operative stress.[3] Symptomatic signs and symptoms, such as tachycardia, diaphoresis, seizures, and weakness, prompt abrupt glucose level evaluation.[3] It is thus recommended to monitor blood sugar regularly, minimize preoperative fasting, or keep the patient on preoperative glucose infusion.[3,4]

RSS patients, particularly infants, are susceptible to hypothermia due to their abnormally great cranial-to-body mass ratio, insufficient subcutaneous fat insulation and small muscle mass, and a large surface-to-weight ratio, which acts as a small insulating mass. This makes the head have the greatest tendency of radiative thermal loss.[3] It is important to be aware of the potential risk of gastroesophageal reflux (GOER) in patients with RSS, which in some situations may require manometry or endoscopy to rule out reflux.[2] In the case of GOER, rapid sequence induction should be done.[4] Our patient had been tested previously with a modified barium swallow, and GOER has been ruled out; hence rapid sequence induction was unnecessary.

Pharmacological implications in patients with RSS center around modifying the dose according to body area rather than body weight.[3] The anesthesia provider must carefully select appropriate drugs and dosages and closely monitor the patient's response to anesthesia to prevent drug toxicity and other complications. Muscle relaxants must be cautiously managed in these patients due to their inconsistent body composition, making neuromuscular blockade monitoring essential.[3,5] Also, it is recommended to be avoided in cases with a suspected difficult airway,[4,7] as was our case. Additionally, individuals with minimal body fat will likely experience a rapid

"wash-out" of volatile anesthetic agents. This can contribute to a quicker recovery from anesthesia due to the rapid release into the circulation.[3] Sevoflurane is the ideal inhalation volatile agent for the pediatric group due to its rapid induction and emergence from anesthesia and low incidence of airway adverse effects. In the presence of cardiac anomalies, administering appropriate prophylactic antibiotics is of utmost importance.[4]

Conclusion

In summary, the anesthetist considered several factors before administering anesthesia to the two-year-old child with Russell-Silver syndrome for elective hip spica surgery. These factors included the patient's medical history, physical examination, vital signs and laboratory values, weight and BMI, and Mallam-Pati classification. The anesthetist carefully reviewed this information to identify potential issues affecting the patient's ability to tolerate anesthesia, anticipate airway management issues, and plan accordingly for safe and successful anesthesia induction and maintenance.

Ethical approval

Bahrain Defense Force Royal Medical Services Research & Research Ethics Committee has given ethical approval with reference number 2022-677.

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Nil.

Conflict of interest

None declared

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