**Título:** PERIOPERATIVE MANAGEMENT OF A PATIENT WITH RUSSELL-SILVER SYNDROME UNDERGOING MAJOR SURGERY: A CASE REPORT

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**Resumo:**

Russell-Silver syndrome (RSS) is a form of dwarfism characterized by intrauterine growth restriction, severe postnatal growth impairment, dysmorphic facial features including mandibular and facial hypoplasia, skeletal malformations and a constellation of endocrine abnormalities (1).

As RSS patients undergoing surgery often pose an anesthetic challenge not frequently encountered in our practice (2), the goal of this case report is to discuss the management of a patient with this condition undergoing orthopedic surgery.

A 20-year-old patient, ASA III, with RSS presents for elective scoliosis correction surgery. His medical history comprised of an interauricular communication, epilepsy, chronic otitis, scoliosis, clino and camptodactyly, macrocephaly and micrognathia with small mouth opening. Preoperatively, aspiration prophylaxis was administered, glucose levels were regularly monitored and meticulous airway evaluation was performed. Intraoperatively, standard ASA monitorization, bispectral index, train-of-four monitorization and two large bore peripheral accesses were placed. A careful awake fiberoptic orotracheal intubation was performed without complications, using lidocaine for topical anesthesia and midazolam for mild sedation. Throughout the surgical procedure, care was taken to ensure normothermia and an adequate anesthesia depth and analgesia with a propofol and remifentanil perfusion. Additionally, triple anti-emetic prophylaxis was administered. The surgery was uneventful and postoperative recovery took place in a post-anesthetic care unit. For postoperative pain management a patient-controlled-analgesia fentanyl perfusion was instituted.

This case report emphasis how, in order to prepare an adequate anaesthetic approach, a deeper understanding of the complexities and potential anaesthetic risks of a disease like RSS is necessary. Patients with RSS have an uncommon body morphology and composition that will often be associated with a difficult airway (3), gastric motility defects and will be more susceptible to hypothermia and hypoglycemia (2). Despite all of these potentially dangerous anaesthetic intercurrences, they may be easily prepared for by a fully informed anaesthesiologist.

Key words: Russell-Silver syndrome; scoliosis correction surgery; perioperative management.

References:

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