CLINICAL VIGNETTE

Anesthetic Management of a Pediatric Patient with Russell-Silver Syndrome

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Background

Russell-Silver Syndrome (RSS) is a growth disorder that leads to slow growth in utero in addition to postnatal growth impairment. Patients typically have failure to thrive (FTT) with normal head growth, leading to a discrepancy between head and body size.[1] Children with this disorder are short and often develop hypoglycemia due to lack of feeding and poor appetite. Other significant impairments include mandibular and facial hypoplasia, with many having a small triangular face, prominent forehead, narrow chin, small jaw and downturned corners of the mouth.[1] In addition, limb asymmetry and endocrine abnormalities can be observed.[2]

The genetic causes of RSS are complex and are believed to be related to mutations in particular genes that control growth. Most mutations are sporadic, but RSS can be inherited in an autosomal dominant or recessive manner. So far, research suspects that close to 50 percent occur as a result of methylation of the short arm of chromosome 11p15 with other abnormalities involving chromosome 7. However, in approximately 40% of patients with RSS, the cause is unknown.[1] Globally, the incidence of RSS range from 1:30,000 to 1:100,000 births.[3]

Case Report

A 17-month old 7.9 kg male with a body mass index (BMI) percentile of 1.23 was electively scheduled for sedation for an MRI of the brain and orbits with and without contrast for a diagnostic evaluation of optic nerve cupping and atrophy. His medical history included a patent foramen ovale, chronic sinusitis, failure to thrive, gastroparesis, gastroesophageal reflux disease (GERD) status post Nissen fundoplication, dysphagia, hypogonadism, laryngomalacia, laryngopharyngeal reflux disease, cyclic neutropenia, plagiocephaly, and volvulus status post repair. Review of systems was negative except medical history that is documented above. The patient had no known drug allergies and was prescribed amoxicillin-clavulanate 400 mg, pediatric multi-vitamin oral liquid and ferrous-iron oral liquid at the time of presentation.

Noteworthy physical exam findings included a low BMI of 13.68 kg/m2 and an axillary temperature of 35.7 degrees Celsius, but physical exam was otherwise normal. Airway exam demonstrated midline trachea, mallampati classification of 1, no damage to dentition, normal mouth opening and adequate thyromental distance. Patient also had no prior anesthetic issues and his past surgical history was significant for a laparoscopic Nissan fundoplication and insertion of a gastrostomy tube at 11 months old. An echocardiogram was significant for an aorto-pulmonary collateral originating from the early descending aorta, a patent foramen ovale and mild left ventricular dilatation.

The patient was scheduled for an MRI of the brain and orbits with and without contrast under monitored anesthesia care. The child's blood glucose level was checked before and after the administration of monitored anesthesia care and was within normal limits. The anesthesia entailed the use of propofol and midazolam. Patient received oxygen supplementation (5L/min) throughout the duration of the anesthesia and the case proceeded uneventfully during anesthesia administration and upon awakening.

Discussion

Russell-Silver Syndrome (RSS) is usually associated with a multitude of phenotypic and metabolic abnormalities that an anesthesiologist should be aware of when taking care of a patient with this disease. Various endocrinopathies have been associated with RSS, the most common being hypoglycemia. The abnormal glucose homeostasis and easily depleted glycogen storages in these patients compounded with prolonged fasting and surgical stress make hypoglycemia a significant risk factor for these patients perioperatively. This is why attention to glucose levels should be made prior to induction. Diaphoresis, tachycardia, seizures, and lightheadedness may indicate hypoglycemia and should prompt immediate evaluation of glucose levels.

A major concern for anesthesiologists regarding patients with RSS is that they are more likely to have a difficult airway due to the facial dysmorphism (hypognathia or retrognathia) and small mouth.[4] The small mouth in a RSS patient may lead to difficulty in mask ventilation and an inadequate mouth opening leading to a challenging direct laryngoscopy. Thus, the anesthesiologist should be prepared to follow the difficult airway management algorithm and have necessary supplies such as a laryngeal mask airway or fiberoptic equipment on standby. In our patient's case, there were no issues with masking the patient and monitored anesthesia care (MAC) was performed.

In addition, special consideration to patient position is needed due to the likelihood of skeletal malformations in RSS. Infants with RSS are also prone to hypothermia more-so than normal infants considering the relatively large circumferential surface area of the head in these patients.[1] Because of this, increased attention should be placed on the vitals of these patients during the procedure. Awareness of potential gastroesophageal reflux (GERD) is mandated in these patients due to higher incidences of this condition in this patient population, and a manometry or endoscopy may sometimes be utilized to rule out reflux. Our patient is status post Nissen fundoplication for his GERD, a swallow study was done and the patient needed no further interventions.

Our patient underwent MRI of the brain and orbits uneventfully under MAC anesthesia and tolerated the procedure well. In the case that a problem arose due to hypoglycemia, hypothermia or difficulty with airway, our anesthetic plan to address these issues would have been implemented due to awareness of the patient's potential anesthetic risks. Our patient continued to do well and was discharged after a short stay in the recovery room.

Conclusion

We present the case of an underweight infant with multiple medical comorbidities and a rare diagnosis of Russell-Silver syndrome who underwent monitored anesthesia care for an MRI of the brain and orbits with and without contrast. He was successfully anesthetized with midazolam and propofol with no issues throughout the duration of the MRI or upon awakening. It can be challenging managing patients with Russell-Silver syndrome and the anesthesia provider should pay close attention to the following areas: airway, hypoglycemia, hypothermia, and patient positioning.

REFERENCES

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Submitted April 10, 2019