Anesthetic Challenge in Corrective Hand Surgery in a Child with Russell–Silver Syndrome

Akshay K Gadre, Kasturi H Bandyopadhyay, Mumtaz Afzal, Amiya K Mishra

ABSTRACT

Introduction: Co-morbidities associated with Russell–Silver syndrome (RSS), viz. severe postnatal growth retardation, congenital heart disease, dysmorphic facial features, limb asymmetry, various patterns of hypogonadism, and constellation of endocrine abnormalities, present significant anesthetic challenge during surgery. We report a case of Russell–Silver syndrome in a 5½-year-old boy who underwent corrective hand surgery and the anesthetic challenges he presented. Russell–Silver syndrome has an autosomal dominant inheritance pattern with variable expressivity and its diagnosis remains primarily clinical, as no definite etiology or specific tests have been established yet. We anticipated difficulty in airway management of this child and so we ensured availability of ancillary equipments and presence of skilled anesthesiologists. This helped us to conduct surgery successfully without any anesthetic complications.

Keywords: Difficult airway, Postnatal growth deficiency, Russell–Silver syndrome.

INTRODUCTION

Russell–Silver syndrome, also called Russell–Silver dwarfism, is a form of dwarfism characterized by intrauterine growth retardation with severe postnatal growth impairment, dysmorphic facial features including mandibular and facial hypoplasia, limb asymmetry, endocrinial abnormalities including hypoglycemia and various patterns of hypogonadism. Russell and Silver were the first to describe this syndrome in 1953 and 1954 respectively. Its incidence ranges from 1 in 3,000 to 1 in 100,000 live births, occurring in all races with equal distribution in males and females.

CASE REPORT

A 5½-year-old boy, weighing 12.5 kg, 100 cm tall, was posted for extensor pollicis longus tendon transfer of left thumb for correction of a congenital malformation. The child was diagnosed with RSS in early infancy, based on low birth weight (2.0 kg), postnatal growth deficiency, characteristic facial features like triangular face with frontal bossing, and hypoplastic mandible (Fig. 1). His height and weight were below the third percentile with delayed developmental milestones and with normal intelligence for his age (Fig. 2). He was active and playful with good effort tolerance. Parents gave history of recurrent respiratory tract infection with an episode of bronchospasm 1 year back which was treated with Salbutamol inhaler for 1 month. He had two episodes of generalized tonic–clonic seizures at the age of 4 years. Since then he had been on treatment with oral valproate.

Physical examination revealed asthenic boy with little muscle mass and scanty subcutaneous fat, narrow elongated head, small triangular face, low-set posterior-rotated ears, hypoplastic mandible, fish-like mouth with down-turned angles, pouted lips with microdontia, and normal genitalia (Fig. 3). Airway examination revealed adequate mouth opening and Mallampati score II predicting ease of tracheal intubation. His left thumb was rudimentary with contracture of left first web space. He was unable to move the thumb due to weakness of dorsiflexor, abductor, and extensor muscles.
Anesthetic Challenge in Corrective Hand Surgery in a Child

Human growth hormone stimulation analysis revealed insufficient basal (0.78 ng/mL) and post-stimulation levels (5.44 ng/mL) after ingestion of 25 μg of Clonidine. Thereafter, he had been on injection of Norditropin 1.3 mg subcutaneously daily till date. Molecular deoxyribonucleic acid analysis was inconclusive of RSS. Examination of cardiovascular system and echocardiography was within normal limits. Chest X-ray showed no abnormalities. Basic hematological and biochemical parameters were within normal limits. There was no hypoglycemia.

Anesthetic Management
The child was shifted to the operating room (OR), unpremedicated, after 8 hours of fasting for solid food, 2 hours for clear liquid, and nebulization with 2.5 mg Salbutamol 1 hour prior to surgery. Standard American Society of Anesthesiologists monitors were attached; intravenous (IV) 1% dextrose in Ringer lactate solution started via 22-gauge IV cannula and injection Glycopyrrolate IV was given as anti-sialagogue agent. Preinduction blood glucose level was 88 mg/dL. Anesthetic plan A was to provide general anesthesia with endotracheal intubation (ETI), while plan B was to secure the airway with ProSeal laryngeal mask airway (PLMA) if the former failed. Difficult airway cart was kept ready prior to induction of anesthesia.

Following preoxygenation, anesthesia was induced with IV injection of Propofol (25 mg). After confirming adequacy of bag-mask ventilation with No. 2 anatomical face mask aided by insertion of size 1 Guedel’s oropharyngeal airway, injection of Succinylcholine 25 mg IV was given to facilitate ETI. Laryngoscopy with a Macintosh size 2 blade revealed Cormack Lehane grade IIIA view. The ETI using 4.5 mm uncuffed endotracheal tube could not be achieved even after two attempts with a gum elastic bougie, hence PLMA no. 2 was inserted with confirmed position and adequate ventilation. Intermittent positive pressure ventilation was done in pressure control mode, and anesthesia was maintained with O₂:air (50:50) + Sevoflurane 1.5 to 2%, injection Fentanyl (25 μg), and titrated doses of injection Atracurium at regular intervals. Multimodal analgesia was administered with injection of Paracetamol (180 mg, IV) and infiltration of incision line with 5 mL of 0.25% Bupivacaine. Patient’s core temperature was measured with esophageal probe and was maintained at 36°C with the aid of warming blankets, maintaining the room temperature at 27°C, humidifying the anesthetic circuit with a moisture exchanger, and using warm IV fluids. Intraoperative blood glucose was measured and found 118 mg/dL. Surgery lasted 3 hours. The child was reversed with neostigmine 0.05 mg/kg and Glycopyrrolate 0.004 mg/kg. He was extubated in the OR after an uneventful anesthetic emergence and recovery. He was monitored for an hour in the postanesthesia care unit. The child had an uncomplicated and smooth recovery and was discharged on the third postoperative day.

DISCUSSION
The RSS is known to have autosomal dominant inheritance pattern with variable expressivity. Maternal uniparental disomy of chromosome 7, in which a child inherits both copies of a region of the chromosome from the mother, has been shown to play a role. However, no single explanation till date has accounted for the heterogeneity of the phenotypic findings. The preanesthetic evaluation and preparation to rule out various abnormalities like congenital heart disease; urogenital, orthopedic, ocular, dental anomalies; and various endocrinopathies is required. Diagnosis of RSS remains clinical as no definite etiology or specific tests have been established. The five core clinical diagnostic criteria are intrauterine growth retardation, poor postnatal growth, preservation of
occipitofrontal circumference, classic facial phenotype, and asymmetry of extremities.

A patient fulfilling any four of these five criteria is diagnosed with RSS.12 Our patient demonstrated four of these clinical criteria. The lack of muscle mass and subcutaneous fat renders these patients more susceptible to hypothermia. Therefore, a thermally neutral operating room, warmed and humidified respiratory gases, drapes, IV fluids, warm irrigating solutions, and wrapping of the head and extremities of the patient should be ensured. Various endocrinopathies like hypopituitarism and adrenocortical insufficiency have been reported in these patients, hypoglycemia being the commonest.13 Hypoglycemia was found to be responsible for symptoms like seizures, tachycardia, excessive sweating, and periodic episodes of severe weakness relieved by food.14 Our patient did not have any history of hypoglycemia but did have history of seizure, the cause of which was not obvious. Pathophysiology of hypoglycemia in RSS can be due to rapid depletion of limited hepatic glycogen stores, or due to unusually large cranial-to-body mass ratio leading to disproportionately increased utilization of glucose, and therefore the possibility of abnormal glucose homeostasis during prolonged fasting and surgical stress should be considered.15 Perioperative glucose status monitoring is must and any unexplained tachycardia, diaphoresis, or excessive somnolence following anesthetic emergence should prompt measurement of blood glucose. Since our patient had history of recurrent upper respiratory tract infection associated with bronchospasm, congenital cardiac anomaly was ruled out.

Obtaining a good mask seal in these patients is likely to be difficult owing to facial asymmetry and small receding chin. Hence, different types and sizes of masks should be available along with airway adjuncts. Facial manifestations of RSS may also render exposure of the vocal cords difficult with conventional laryngoscopy. Since difficult intubation was anticipated, adequate size PLMA was kept ready, which eventually helped us to secure the airway. A pediatric fiberoptic bronchoscope of 3.1 mm external diameter should also be handy. Preoperative administration of anti-sialagogue agent helps in decreasing secretions in the airways and aids visibility.16

Since these patients have underdeveloped muscle mass, the recommended dose of any neuromuscular blocking agent may cause profound relaxation. Thus, careful titration of top-up doses along with neuromuscular blockade monitoring is advisable. Recovery from inhalational anesthetic agent is usually quick, which was also observed in our patient. The “wash-out” of volatile agents occurs at a rapid rate due to the lack of significant fat for uptake during maintenance of anesthesia and the subsequent release into the circulation during emergence. At the end of surgery, the boy was adequately reversed and extubated in the operating room.

CONCLUSION

We have presented the anesthetic implications of RSS along with our experience of managing such a case. We encountered management concerns with regard to mask ventilation, laryngoscopy, and ETI for which we were forearmed. Temperature regulation, dosing of neuromuscular blocking agents, glucose homeostasis, and emergence from anesthesia need to be managed with particular care. Awareness of concomitant anomalies associated with RSS, such as congenital heart disease, hypopituitarism, and adrenal insufficiency, is essential for proper care.

REFERENCES