



Management of a Parturient with Russell Silver Syndrome, Cerebral Palsy, and HELLP Syndrome

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Background

Russell Silver syndrome is a rare congenital disorder associated with:

- Dwarfism
- dysmorphic facial features
- intrauterine growth restriction
- postnatal growth impairment
- endocrine abnormalities

Comprehensive preoperative assessment and preparation is required in order to avoid anesthetic and obstetric complications in RSS patients.

Clinical Presentation

A 28-year-old G1P0 at 26 weeks with a past medical history of Russell-Silver Syndrome (RSS) and cerebral palsy presented to the L&D unit for short cervix and preterm contractions, found to have clinical evidence of preeclampsia.

Features of her RSS/Cerebral Palsy

Short stature (4 ft, 33 kg)
Hypoplastic mandible
Growth deficiency
Bilateral lower extremity contractures/weakness
wheelchair use for mobilization

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Clinical Course

Patient presented to labor and delivery for evaluation of preterm labor after a routine clinic exam revealed short cervix and preterm contractions.

Her laboratory findings were significant for

Protein/creatinine ratio 0.45

Platelet 64

Uric Acid 8.1

New-onset pulmonary edema, worsening thrombocytopenia, and transaminitis raised concerns for HELLP syndrome, prompting urgent cesarean delivery.

General endotracheal anesthesia was required for avoidance of epidural hematoma risk with thrombocytopenic coagulopathy



Figure 1: Radiographic films in evaluation of skeletal dysplasia



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Clinical Course (Con't)

- Prudent airway management with video laryngoscope and rapid sequence induction with rocuronium.
- Management included judicious medication administration related to patient size, as well as recognition of endocrine/metabolic associated complications of RSS.
- The patient made an uneventful recovery and was discharged on postoperative day 5
- The premature neonate delivered with APGAR Scores of 4-9 was admitted to the neonatal ICU.

Discussion

Russell Silver syndrome (RSS) is a rare congenital disorder associated with dwarfism, dysmorphic facial features, intrauterine growth restriction, postnatal growth impairment, and endocrine abnormalities.

Preparation includes recognition of the following:

- Facial and laryngeal anatomic alterations that can contribute to airway management challenges
- Nutritional deficiencies and endocrinologic complications (hypoglycemia)
- Challenges related to neuraxial anesthesia-difficult patient positioning, level of termination of conus, spinal deformities, muscle spasms
- Recognition of the risk of requirement for cesarean delivery in the setting of cephalopelvic disproportion.

References

1. Scarlett MD, Tha MW. Russell-Silver syndrome: anaesthetic implications and management. West Indian Med J. 2006;55:127-129
2. Dinner M, Goldin EZ, Ward R, Levy J. Russell-Silver syndrome: anesthetic implications. Anesth. Analg. 1994;78:1197-1199.
3. Spiteri BS, Stafrace Y, Calleja-Agius J. Silver-Russell Syndrome: A Review. Neonatal Netw. 2017 Jul 1;36(4):206-212.