

Sickle Cell Disease Complicated by Difficult Pain Management and Additional Hgb Mutations

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● **Patient History**

- 19-year-old G1P0 admitted for IOL at 37 weeks gestation in the setting of fetal anemia
- **Past medical history:**
 - ▶ Sickle cell disease (SCD) on hydroxyurea
 - Childhood diagnosis complicated by acute chest syndrome (2015, 2016, 2022)
 - Recurrent pain crises requiring regular admissions, but none during this pregnancy
 - Concurrent gamma globin mutation in Hgb G2 gene (required for fetal Hgb)
 - Low SpO2 ~86-94%, seen by pulmonology without definitive explanation
 - ▶ Asthma – well-controlled on inhalers
 - ▶ Pregnancy complicated by fetal anemia requiring multiple IUTs (Intrauterine fetal transfusion)

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● Hospital Course

• Antepartum Management

- ▶ Frequent IUTs complicated by anxiety, requiring GA or sedation

• Intrapartum Pain Management

- ▶ Early epidural complicated by multiple failed and dislodged catheters requiring 4 total placements
- ▶ Failed induction of labor on day 4
- ▶ Cesarean delivery under spinal anesthesia and sedation

• Intrapartum Complications

- ▶ Increased risk of methemoglobinemia requiring cautious use of specific medications including local anesthetics
- ▶ Supplemental O₂ via NC or facemask due to low SpO₂
 - Monitoring with frequent ABGs

HEMOGLOBIN ELECT...	
Hemoglobin A	22.2 ↓
Hemoglobin A2	2.7
Hemoglobin F	9.4 ↑
Hemoglobin S	65.7 ↑
Hemoglobinopathy Eval	Abnor... !

BLOOD GAS / COOX / ...	
Blood Gas Source	Arterial
pH, Blood	7.41
PCO2	31 ↓
Oxygen Content	10.9 ↓
PO2	104
Oxygen Saturation	98
Bicarbonate	20 ↓
Base excess	Neg 5.0
Hemoglobin, Whole Blood	8.1 ↓
Oxyhemoglobin	93.8
Carboxyhemoglobin	2.9 ↑
Methemoglobin	1.6 ↑
FIO2	Not specified

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● Discussion

- Increased risks associated with pregnancy in SCD patients, including:
 - ▶ Vaso-occlusive episodes, severe anemia, endothelial dysfunction, hypercoagulable state and high metabolic demands
- Acute chest syndrome is one of the most common causes of death
- Increased risk of Cesarean section (up to 43% in the nulliparous group)
- Pulse oximetry readings may not correlate with PaO₂ in the SCD population or patients with known additional hemoglobinopathies
- Neuraxial local anesthesia was tolerated in this case without clinically significant increases in MetHgb levels

● Key Points

- Pain control is a priority for adequate care in the SCD population
 - ▶ Neuraxial anesthesia is the gold standard
- Patients with SCD warrant increased attention and multidisciplinary management c/o hematology, obstetric, and anesthesia providers