Crisis Averted: A Multidisciplinary Approach to Sickle Cell Disease in Pregnancy

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Introduction

- Sickle cell disease (SCD) is an inherited hemoglobinopathy characterized by abnormal hemoglobin polymerization, affecting nearly 8 million individuals globally
- Common triggers of acute crises include: dehydration, infections, cold temperatures, stress, and low oxygen levels
- Though a majority of SCD pregnancies result in favorable outcomes, SCD in pregnancy carries increased risks of vaso-occlusion, infection, and maternal and fetal adverse events
- Here we present a case of premature Cesarean delivery (CD) in a SCD pregnancy from a multidisciplinary perspective

Maternal/Fetal Complication	Incidence in SCD	Control
Pre-eclampsia	9.7-15.7%	3.2-6.2%
Urinary tract infection	7.1-7.9%	0.6-3.4%
Acute Chest Syndrome	22.1%	1.7%
Maternal Mortality	0.5-3.6%	~0%
Low birth weight	18.6-46%	9.1-19%
Spontaneous abortion	34.4%	10.8%
Stillbirth	4.8-8.6%	0.8-1.9%
Neonatal death	1.1-2.6%	0.9-1%

Table 1. Incidence of select adverse events in SCD pregnancies. Information from references 3 and 4

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Case Presentation

One-liner: A 33-year-old G2P0100 at 33 weeks 2 days gestation, PMH significant for asthma and SCD complicated by prior acute chest syndrome, pulmonary hypertension, iron-overload-related myometrial damage, extensive alloimmunization, and prior intrauterine fetal demise secondary to hemolytic transfusion reaction admitted for acute pain crisis.

ADMISSION

- Extensive alloimmunization
- Prior IUFD secondary to hemolytic transfusion reaction
- Admitted in setting of acute pain crisis

ANTEPARTUM

- Planned for external cephalic version but self converted
- H/H 6.5/19.8%, transfused with 1 unit of crossmatched pRBCs prior to IOL

LABOR

- IOL w/ aROM at 34w5d under CSE
- C/b chorioamnionitis and failure to progress prompting category 2 cesarean delivery

INTRA-OP

- Standard ASA monitors + arterial line + 2 18g PIVs + O2 via NC
- Existing epidural dosed up w/ 2% lido
- Intra-op Abx: azithromycin, gentamicin, clindamycin
- Uncomplicated delivery

POST-OP

- Epidural removed post-op
- Fever and tachycardia on POD2
- Empiric abx started
- Negative hemolysis labs
- Pain control inadequate, treated with dPCA
- Discharged POD7

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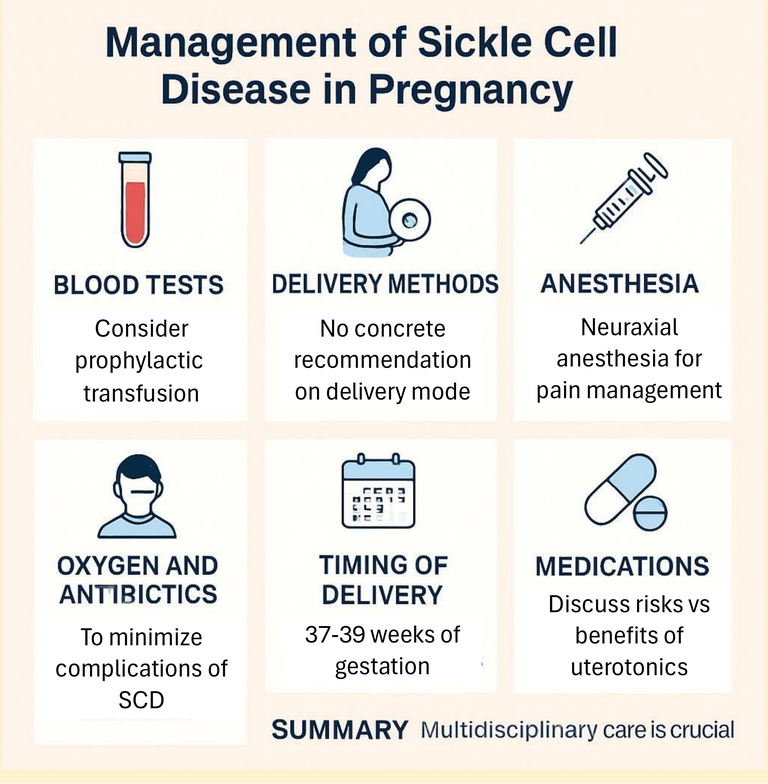
Discussion & Key Points



- Planning for labor and delivery in SCD pregnancy involves blood tests such as blood typing and cross matching and consideration of prophylactic transfusion
- There is no concrete recommendation for mode of delivery and decision to undergo CD is guided by standard obstetric indications
- Neuraxial anesthesia avoids airway manipulation and has superior pain control and minimizes risk of bleeding compared to general anesthesia
- Oxygen supplementation reduces development of acute chest syndrome
- Standard prophylactic antibiotics to reduce increased risk of infections SCD patients face in pregnancy
- IOL/CD most efficacious at 37-39 weeks of gestation rather than at full term to mitigate the risks of high rates of placental insufficiency, maternal vaso-occlusive crisis (VOC), and stillbirth

References

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