



How low can you go? An Emergency Caesarean section in a patient with Sick Cell disease and Hyperhaemolysis Syndrome

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Introduction

Sickle Cell Disease (SCD) in pregnancy is associated with increased morbidity and mortality due to physiological changes that exacerbate pre-existing organ dysfunction.

Patients with SCD, due to increased transfusion requirements, are at higher risk for transfusion-related complications such as delayed haemolytic transfusion reactions (DHTR) and the rare but serious Hyperhaemolysis Syndrome (HHS).

HHS occurs when intravascular haemolysis affects both transfused and autologous red blood cells, resulting in haemoglobin (Hb) levels below the pre-transfusion range (1).

Further transfusions can escalate the problem, and cell salvage is contraindicated, making HHS in pregnancy a significant challenge for anaesthetists during the perioperative and peripartum periods.

	Delayed Haemolytic Transfusion Reaction (DHTR)	Hyperhaemolysis Syndrome (HHS)
Severity	Mild to severe	Severe
RBC destruction	Transfused RBCs	Both transfused and patient's own RBCs
Haemoglobin Level	Static or inadequate rise	Drop to below transfusion levels
Cause	Alloimmunisation	Can occur in absence of new antibodies
Direct antiglobin test (DAT)	Positive	Negative (or faint +ve)
Haemoglobinuria	Rarely	Common

Case Report

- 40-year-old primigravida with known SCD, baseline Hb of 82-84 g/dL, and no prior transfusion complications.
- At 34+4 weeks, the patient was admitted with low oxygen saturations but remained asymptomatic with a reassuring CTG.
- She received one unit of packed red cells and was discharged 48 hours later
- Two weeks later, presents to a district hospital with fever, low saturations, Hb 69g/dL, and dark urine.
- DHTR was suspected and treatment promptly initiated
- Hb continued to fall to 47g/dL and she was transferred to a tertiary hospital ICU.
- HHS was diagnosed, and no further transfusions were permitted. She was treated with Eculizumab, IV methylprednisolone, folic acid, and EPO.
- After Multi-Disciplinary Team (MDT) discussion, decision was made to expedite delivery due to ongoing haemolysis and concerns about fetal oxygen delivery.
- A caesarean section was performed without complication at 37+1 weeks under Combined Spinal Epidural (CSE), arterial line. EBL: 1L
- Hb dropped to 37g/dL on day one postoperatively before beginning to recover.
- Mother and baby were discharged home on day 12 post delivery

Discussion

We describe a rare case of HHS in a primigravida with SCD and notable anaemic tolerance.

Haemoglobin levels pre and post delivery are below any previously reported for a caesarean section with active haemolysis. Due to prompt diagnosis and multidisciplinary management, both mother and baby had an excellent outcome.

HHS is a rare but potentially fatal condition in pregnancy. This case highlights the importance of:

- Early diagnosis (symptoms can mimic a vaso-occlusive crisis)
- Specialist input at an early stage of management
- Timely intervention
- MDT approach to diagnosis and management

References

1. Madu AJ, Ugwu AO, Efoji C. Hyperhaemolytic syndrome in sickle cell disease: clearing the cobwebs. Medical Principles and Practice. 2021; 30(3): 236-43.

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