

Transfusion Camp Rwanda May 2024

“Sickle Cell Disease and Transfusion”, developed by Drs. Jacob Pendergrast, Lianne Rotin and Aggrey Dhabangi

Case 1

A 30 year-old female with HgbS β^0 presents to the emergency department with acute onset pain in her lower back and shins, consistent with her usual vaso-occlusive pain crises. She is under shared care at your institution and a local peripheral hospital closer to her home. On examination she is alert and oriented, with all vital signs within normal limits with the exception of sinus tachycardia. Physical examination reveals conjunctival pallor, scleral icterus, and digital clubbing. She is initially managed with intravenous (IV) fluids, supplemental oxygen and frequent doses of morphine sulfate, but after eight hours there is little improvement in her pain symptoms. Her current laboratory investigations reveal a hemoglobin (Hgb) of 63 g/L, white blood cell count (WBC) $8.1 \times 10^9/L$, and platelets $225 \times 10^9/L$. Her reticulocyte count is elevated at $200 \times 10^9/L$.

1. What is the next best step in her management?
 - a) MRI scan of lumbar spine
 - b) Obtain transfusion history from other hospitals where she has been treated
 - c) Presence of anti-Parvovirus B19 antibodies
 - d) Transfuse 1 unit of RBCs

2. Given the patient’s current status, what should the goal of transfusion therapy be?
 - a) Decrease HgbS to < 30%
 - b) Maintain Hgb > 50 g/L
 - c) Target Hgb at approximately 100 g/L
 - d) Transfuse with the goal of normalizing physiologic targets such as serum lactate, troponin and creatinine

3. When selecting blood products for patients with sickle cell disease, it is most important that they be:
 - a) As fresh as possible
 - b) Matched for RhCE and K
 - c) Sickledex[®]-negative
 - d) Irradiated

Case 2

An otherwise previously-well 14 month-old baby girl with HgbSS is brought to the emergency room by her mother after the baby was noted to be increasingly irritable, with pallor, jaundice, and thready pulses. On examination, the baby is confirmed to be pale and icteric, with a tender mass palpable at the umbilicus. Heart rate is 170 bpm, with blood pressure of 70/40 mmHg. The baby is afebrile with pulse oximetry of 100% SpO₂ on room air. Laboratory investigations reveal a Hgb of 52 g/L, a WBC of $3.4 \times 10^9/L$, a platelet count of $100 \times 10^9/L$, with normal coagulation times but serum chemistry revealing a newly elevated creatinine at 130 $\mu\text{mol/L}$. You diagnose the patient with splenic sequestration crisis.

4. How will you manage this patient?
 - a) Give isotonic fluids and vasopressors
 - b) Initiate exchange transfusion to target HgbS <30%
 - c) Provide simple pRBC transfusion (3-5 cc/kg)
 - d) Send the patient for urgent splenectomy

5. Which transfusion reaction is this patient most at risk for?
 - a) Delayed hemolytic transfusion reaction
 - b) Hyperkalemia
 - c) Hyperviscosity
 - d) Transfusion-associated circulatory overload

Case 3

A 17 year-old man with HgbSC is recovering on the ward after undergoing a right total hip replacement for avascular necrosis. On post-op day #2 he begins experiencing chest pain, fever and dyspnea. Physical examination reveals a patient in moderate respiratory distress but is alert and oriented, with HR 80 bpm, BP 110/70, RR 24, Temp 38.2°C and SpO₂ of 95% on 2L O₂ by nasal prongs. He has mild conjunctival pallor and scleral icterus, with bilateral inspiratory crepitations. Jugular venous pressure (JVP) is 2 cm above sternal angle and there is no peripheral edema. Laboratory investigations reveal a Hgb of 80 g/L (100 g/L pre-op), WBC of $16 \times 10^9/L$ (6), and a platelet count of $400 \times 10^9/L$ (250), with normal coagulation times and serum chemistry, and BNP, troponin and serum lactate levels all within normal limits.

6. Which one of the following features on CXR would suggest the patient is experiencing an acute chest syndrome?
 - a) Decreased lung volumes
 - b) Enlarged pulmonary arteries
 - c) Interstitial edema
 - d) Lobar consolidation

7. What sort of transfusion support should you provide this patient?
 - a) None
 - b) Exchange RBC transfusion
 - c) Therapeutic phlebotomy
 - d) Top-up RBC transfusion

8. Which one of the following interventions could have prevented this complication?
 - a) Aggressive hydration at 1.5x maintenance for first 48 hours post-op
 - b) Full anticoagulation as soon as surgical hemostasis attained
 - c) Hydroxyurea
 - d) Pre-operative exchange transfusion

Case 4

A 28 year-old woman with HgbS β ⁺ is admitted to hospital with a hemorrhagic ovarian cyst and a Hgb of 60 g/L (baseline 95 g/L). She is transfused 2 units of RBCs and undergoes an otherwise uncomplicated oophorectomy. One week after discharge she presents with a vaso-occlusive pain crisis. Her initial Hgb is 78 g/L but over the course of 48 hours it falls to 59 g/L, accompanied by a stable WBC of 12×10^9 /L and platelet count of 180×10^9 /L. She is hemodynamically stable and abdominal imaging confirms no ongoing bleeding or hepatosplenomegaly, but her LDH increases to 850 U/L, with indirect bilirubin of 50 μ mol/L. Reticulocyte count decreases from a baseline of 400×10^9 /L. She is transfused 1 unit of RBCs; the blood bank notifies you that the pre-transfusion sample reveals an anti-E antibody that was not detectable on her earlier sample from 1 week ago. Direct antiglobulin test is negative. The next morning after her transfusion her Hgb is 50 g/L. She is transfused another unit of RBCs and her Hgb the next morning has fallen again to 42 g/L. The patient remains hemodynamically stable but is complaining of increasing fatigue.

9. What is the most likely explanation for the lack of response to transfusion?
 - a) Autoimmune hemolysis
 - b) Hyperhemolysis
 - c) Intra-abdominal bleeding
 - d) Units are serologically incompatible (ie., delayed hemolytic transfusion reaction)

10. What is the first-line treatment for this patient?
 - a) Eculizumab and rituximab
 - b) Intravenous iron
 - c) IVIG and steroids
 - d) Top-up transfusion to Hgb > 80 g/L, followed immediately by exchange transfusion

Case 5

A 31 year-old woman with HgbSS is admitted to obstetric triage with a generalized pain episode. Of note, she had been taking hydroxyurea with excellent disease control prior to becoming pregnant, but has held it due to concerns with potential teratogenicity. Her Hgb at her current presentation is 65 g/L (baseline 80-90 g/L pre-pregnancy). She is 17 weeks gestation and this is her third acute pain episode in the pregnancy.

11. Which of the following is true about transfusion support during pregnancy in patients with sickle cell disease?
- a) Intrauterine transfusion support may be required to protect the fetus from experiencing vaso-occlusive episodes
 - b) Pregnant women with sickle cell disease should be routinely transfused to maintain their HgbS < 30% in order to optimize fetal development
 - c) Regular transfusion support during pregnancy can decrease maternal pain crises
 - d) Sickling complications tend to be less frequent due to the increase in maternal HgbF% that accompanies pregnancy

TRANSFUSION CAMP RESOURCES ARE DEVELOPED BY TRANSFUSION CAMP FACULTY FOR EDUCATIONAL PURPOSES ONLY. THE RESOURCES **MUST NOT BE USED OR DISTRIBUTED OUTSIDE OF TRANSFUSION CAMP** WITHOUT THE CONSENT OF THE TRANSFUSION CAMP ORGANIZERS. THE MATERIALS ARE NOT INTENDED TO BE A SUBSTITUTE FOR THE ADVICE OF A PHYSICIAN AND SHOULD BE ASSESSED IN THE CONTEXT OF THE APPLICABLE MEDICAL, LEGAL AND ETHICAL REQUIREMENTS IN ANY INDIVIDUAL CASE.

PROVIDE FEEDBACK ON TRANSFUSION CAMP RESOURCES OR ENQUIRE ABOUT TRANSFUSION CAMP BY CONTACTING TRANSFUSIONCAMP@BLOOD.CA.