Transfusion Camp 2022-2023

Day 2: Seminar 2B, November 18, 2022

"Sickle Cell Disease and Transfusion", developed by Dr. Jacob Pendergrast

Case 1

A 30 year-old female with HgbSß^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 x 10^9/L, and platelets 225 x 10^9/L. Her reticulocyte count is elevated at 200 x 10^9/L..

1. Which of the following pieces of information would be most useful in her initial management?
   a) Coagulation times
   b) MRI scan of lumbar spine
   c) Presence of anti-Parvovirus B19 antibodies
   d) Transfusion history from other hospitals where she has been treated

2. Given the patient's current status, what should the goal of transfusion therapy be?
   a) Decrease HgbS to < 30%
   b) Keep serum lactate within normal range
   c) Maintain Hgb > 50 g/L
   d) Target Hgb at approximately 100 g/L

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) Washed

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 x 10^9/L, a platelet count of 100 x 10^9/L, with normal coagulation times but serum chemistry revealing a newly elevated creatinine at 130 µmol/L.

4. Which one of the following statements is true regarding splenic sequestration crises?
   a) They are the most common cause of death amongst children with sickle cell disease
   b) Sickle cell patients undergo autosplenectomy as a complication of sequestration crises
   c) Once a child has had one sequestration crisis, they are likely to have more
   d) Most cases of sequestration crises occur abruptly, without any apparent preceding illness

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 38.2ºC 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 x 10^9/L (6), and a platelet count of 400 x 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) Top-up RBC transfusion
   c) Exchange RBC transfusion
   d) Therapeutic phlebotomy

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 HgbSß$^{+}$ 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 x 10^9/L and platelet count of 180 x 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 µmol/L. Reticulocyte count decreases from a baseline of 400 to 100 x 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) Regular transfusion support during pregnancy can decrease maternal pain crises
    b) Intrauterine transfusion support may be required to protect the fetus from experiencing vaso-occlusive episodes
    c) Pregnant women with sickle cell disease should be routinely transfused to maintain their HgbS < 30% in order to optimize fetal development
    d) Sickling complications tend to be less frequent due to the increase in maternal HgbF% that accompanies pregnancy