Dr. Jacob Pendergrast – Transfusion Support for Sickle Cell Disease

The "Rules" of transfusion differ significantly in sickle cell disease vs. other conditions

- The benefit of transfusion in sickle cell disease comes more from decreasing the blood viscosity (ie., decreasing the HgbS%) than it does with increasing the oxygen carrying capacity (ie., the total Hgb)
- Because the viscosity of sickle cell blood is so high, transfusion may cause harm if it drives the total hemoglobin higher than 100 g/L, and in the absence of acute organ dysfunction is usually not required until the Hgb is less than 50 g/L

Even with severe anemia, transfusion should be pursued cautiously

- The conditions that can cause a patient with sickle cell disease to have a hemoglobin less than 50 g/L also increase the risk of adverse transfusion reactions
    - Aplastic crisis: *hypoxia due to volume overload*
    - Sequestration: *hyperviscosity due to autotransfusion*
    - Hyperhemolysis: *worsening anemia due to bystander hemolysis*

Most sickle cell patients requiring surgery should have a pre-op transfusion, but the type of transfusion will vary

- Transfusions are usually not needed for low-risk patient (ie., no chronic organ dysfunction) with low risk procedure (ie., procedures on distal extremities or perineum)
- For everyone else, the choice of top-up vs exchange transfusion depends on patient comorbidity, their baseline hemoglobin, and whether the procedure is high-risk (ie., requiring a prolonged recovery period)

There is only weak evidence to guide transfusion support in pregnant sickle cell patients

- Available evidence suggests transfusion is of more benefit for maternal well-being than the developing fetus, and therefore should be prescribed in absence of significant maternal symptoms
- There may be exceptions, however (eg., signs of placental insufficiency, previous history of intra-uterine growth retardation)

There is a strong evidence for the benefit of transfusion to prevent stroke

- Transfusion indicated for all children with high-risk transcranial doppler ultrasound and those with a history of symptomatic stroke; there is smaller value for children with silent cerebral infarcts, and transfusion decisions should be made for them on a case-by-case basis
- There is limited evidence to inform transfusion for stroke prevention in adults, and causes other than sickle cell disease should be sought
- Exchange transfusion should be initiated immediately in the setting of acute ischemic stroke, but should wait until bleeding has stopped in patients with hemorrhagic stroke

Therapeutic transfusion is indicated for acute organ compromise but not uncomplicated vaso-occlusive crisis

- Although little evidence, consensus supports transfusion as first line therapy for for acute stroke, acute chest syndrome, and sickle hepatopathy
- Other situations (eg., malleolar ulcers, pulmonary hypertension), transfusion can be considered if other treatments have failed

Selection of RBCs must be done with care

- Sickle cell patients are at higher risk of alloimmunization, and delayed hemolytic transfusion reactions can be deadly
- Extended antigen matching therefore very important, which requires ensuring the blood bank knows your patient has sickle cell disease and their full transfusion history