Guidelines for Surgery in Sickle Cell Disease Patients

**SS and Sβ⁰ Thalassemia:**

**Pre-operative:**
1. Simple transfusion to obtain a **Hb of 10 gm/dl** (Goal: HbS~60%) at the time of surgery  
   a) Sickle negative blood, leukocyte depleted blood  
   b) Antigen matched blood (C,D,E and Kell)
2. Exchange transfusion: Hb S <30%: arrange for red cell pheresis with Blood Bank for the following surgeries:  
   a) Neurosurgery  
   b) Retinal surgery  
   c) Cardiac surgery  
   d) Organ transplants
3. Admission for at least 12 hours of preoperative hydration (1.5x maintenance when NPO)

**SC and Sβ⁺ Thalassemia:**

Patients with Hb SC and Sβ⁺ Thalassemia may not need pre-op transfusions for all surgical procedures. The decision to transfuse is based on type of surgery, length of anesthesia and prior SCD complications of patient.

Transfusions should be done for the following:  
1. Intra-abdominal surgery  
2. Surgeries which may compromise the airway  
3. Orthopedic surgery with tourniquet application  
4. Neuro, cardiac, retinal surgeries, organ transplants  
5. History of significant complications of their disease (particularly ACS)

**Pre-operative:**

1. Simple transfusion to obtain a Hb of 10-11 gm/dl at the time of surgery  
   a) Sickle negative blood, leukocyte depleted blood  
   b) Antigen matched blood (CDE and Kell)
2. Partial exchange transfusion may be required to keep the post transfusion Hb < 11 gm/dl  
3. Admission for at least 12 hours of preoperative hydration (1.5x maintenance when NPO).

**Postoperative Plan:**

1. Continue hydration at 1.5x maintenance  
   a. Initially, IV after procedure  
   b. PO + IV when awake and able to tolerate PO fluids (to equal 1.5x maintenance)
2. 35% oxygen by facemask for 24 hours regardless of sats. Maintain an oxygen saturation > 94%. (This is recommended for all patients with HbSS and HbS beta thal 0 undergoing general anesthesia and patients with Hb SC, HbSbeta thal + who are either undergoing major surgeries, T and A or have prior history of acute chest syndrome).  
3. Monitor with pulse oximetry for at least 6 hours post surgery  
4. Incentive spirometry for a minimum of 2 days post operative if inpatient.  
5. Appropriate analgesia to allow incentive spirometry

(Written/updated by Arpan Sinha, MD and Janna Journeycake MD – June 2019)