1.0 Background

Children with Sickle Cell Disease are at risk of developing post-operative Acute Chest Syndrome. With improvements in intra-operative monitoring and more awareness of the conditions that induce red cell sickling (hypoxia, hypothermia, acidosis, and dehydration), dramatic reductions in perioperative complications have occurred.

It has been shown that the correction of anemia and reduction in the percentage of haemoglobin S will prevent intra-operative and post-operative morbidity and mortality in sickle cell patients. Historically, uncertainty has existed as to the benefits of simple pre-operative transfusion, given the concern of increased blood viscosity. While a partial exchange transfusion would allow for a lowering of hemoglobin S without an increase in hematocrit, a multicenter randomized trial comparing simple and exchange transfusion to prevent peri-operative complications in patients with Sickle Cell Anemia favored simple transfusions for pre-operative management.

In 2011, the Transfusion Alternatives Preoperatively in Sickle Cell (TAPS) study, a multicenter randomized study of transfusion vs. no transfusion pre-operatively, revealed more serious complications amongst patients who had not been transfused compared with those who received a transfusion. The significance of the results prompted premature closure of the trial in order to protect patient safety.

In weighing the risks and benefits of pre-operative transfusion, the extent of the operative procedure, including post-operative dysfunction and pain, must be assessed. A retrospective review by Griffin and Buchanan showed that for the majority of minor elective procedures (hernia repair, circumcision, tympanostomy tube placement, strabismus surgery, and dental rehabilitation) in sickle cell patients, pre-operative transfusions are unnecessary, as these patients usually have uncomplicated courses. Surgeries that place patients with Sickle Cell Disease at higher risk (50%) of developing post-operative complications include thoracotomy, laparotomy, and tonsillectomy/adenoidectomy (T/A). Patients undergoing these and other procedures, characterized by longer intra-operative duration and by compromised chest wall and pulmonary mechanics, may benefit from pre-operative transfusion.

In conclusion, patients who are seriously ill, hematologically compromised (Hgb 15g/L < baseline), or undergoing major surgeries (e.g. thoracotomy, laparotomy), should receive a pre-operative simple blood transfusion. Patients with a history of pulmonary disease or frequent recurrent painful crises requiring hospitalization appear to be at a higher risk of complications, and hence should also be transfused. Patients who are in their usual state of health, at baseline Hgb, and well-established on Hydroxyurea likely do not need a pre-operative transfusion for relatively simple surgeries (cholecystectomy, splenectomy). The decision regarding pre-operative transfusion should be based on the unique past history and current medical condition of the individual patient.
2.0 Pathway

**Sickle Cell Peri-operative Management Care Pathway**

**PRE-OPERATIVE**
- **GOALS**
  - Complete thorough examination.
  - Check that child does not have any acute illness.
  - Position procedure if child is not healthy.
  - Consider transfusion for the following children:
    - Under 88 kg, as a baseline, discuss with Hematology Team.
    - Under 1.8 months or 0.5 kg, discuss with Hematology Team.
  - Plan general anesthetic carefully for elective procedures, in collaboration with Sickle Cell Team.
  - Consider stroke risk and previous episodes of Acute Chest Syndrome as these can lead to post-op complications.
  - Discuss any issues with Hematology/nurse/physio/ICU.
- **PRE-OPERATIVE IN-PATIENT AND INTRA-OPERATIVE**
  - Avoid hypothermia, hypoxia, and hypotension.
  - Ensure child is awake, ventilating, and oxygenating well prior to extubating.
  - Monitor O₂ saturation closely, ensure 92%.
  - In the recovery ward, assesses thoroughly before transferring to in-patient unit.
  - Monitor O₂ saturation closely, ensure 92%.
  - Monitor O₂ saturation closely, ensure 92%.
- **POST-OPERATIVE MANAGEMENT**
  - Pulmonary toilet.
  - Avoid hypo/ hypoxia, acidosis, and/or base.
  - Keep child warm.
  - Ensure adequate pain control.

**PRE-OPERATIVE**
- **GOALS**
  - Admit all patients 1 day prior to surgery.
  - Assess for risk factors and risk of potential complications.
  - Child/children are admitted on-site on arrival. Refer to procedure document.

**PRE-OPERATIVE IN-PATIENT AND INTRA-OPERATIVE**
- Request Hematology and Anesthesia consults.
- Complete pre-operative checklist:
  - Ensure blood group and red cell phenotype are on patient's chart.
  - If surgery is expected to need a blood transfusion pre-op or intra-op, ensure there is a current sample for type and screen (indicated Sickle Cell Disease as diagnosis).
  - Ensure that blood required and Sickle Cell Disease are indicated on OR list.
  - If transfusion ordered pre-op, ensure the indication for the ordering of blood products is indicated Sickle Cell Disease in addition to Transfusion.
  - No contraindications, start incentive spirometer and refer to physiotherapy. See Cardiopulmonary Physiotherapy policy.
  - Ensure child is well oxygenated pre-operatively,apih O₂ at 8L/min when patient is called to OR (initiate O₂ at 15 minutes prior to OR and continue O₂ on route to OR).
  - Ensure child is warm. Avoid warming blankets (from warming device).
- **Anesthetic Management:**
  - Pre-warm O₂ and/or use forced-air warming i.e. Bair Hugger to ensure normothermia and prevent acidosis due to hypothermia.
  - Ensure child is well oxygenated pre-operatively.
  - Induction and intubation should be undertaken with little or no hypotension.
  - Monitor O₂ closely.
- **POST-OPERATIVE MANAGEMENT**
  - Pulmonary toilet.
  - Avoid hypo/ hypoxia, acidosis, and/or base.
  - Keep child warm.
  - Ensure adequate pain control.

**PRE-OPERATIVE**
- **GOALS**
  - Review NPO requirement prior to surgery.
- **PRE-OPERATIVE IN-PATIENT AND INTRA-OPERATIVE**
  - IV and PO fluids at maintenance.
  - Maintain NPO status.
  - When NPO administer IV fluids at maintenance.
  - Refer to Fluid and Electrolyte Guidelines.
- **POST-OPERATIVE MANAGEMENT**
  - Complete serum electrolytes (Na⁺, K⁺, glucose, urine, creatinine) prior to IV fluid administration.
  - IV and PO fluids at maintenance.
  - Use only warm OR room temperature fluids cold co.
  - Maintain hydration to prevent re-oxygenation, hyper-perfusion, and microvascular occlusion. (Its post-administer fluid)
  - Avoid over-hydration. It could result in pulmonary interstitial oedema which can lead to hypoxia and acidosis.
  - Provide appropriate analgesia to ensure that child participates in ambulation and pulmonary clearing.

3.0 References


**Reviewers**

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**Attachments:**

*Revision History.docx*

*SCD Periap 2021 FINAL.pdf*