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 Clinical Practice Recommendations

Statement of Evidence: Recommendations were made by expert group consensus. (Grade C)

2.1 Pre-operative Assessment

2.1.1 Examine the patient thoroughly on pre-op admission to ensure that the child is in good health and without any current acute illnesses. If the child is not in his/her usual state of health, the procedure should be postponed. Consider transfusion for children:

- a. undergoing major procedures—thoracotomy, laparotomy, T/A
- b. who are ill and in whom surgery cannot be postponed
- c. with haemoglobin 15g/L below baseline levels
- d. with significant history of pulmonary disease, stroke, etc.

These issues must be discussed in consultation with a Haematology Fellow / Staff.

General anesthetic needs to be planned carefully for elective procedures in collaboration with the Sickle Cell Team. Consider stroke risk, previous episodes of Acute Chest Syndrome etc. as these can lead to post-operative complications.

2.2 Inpatient Unit Pre-operative Management

2.2.1 All patients should be admitted on the day prior to operation and given IV fluids for a minimum of 8 hours at a total fluid intake (TFI) of maintenance flow rate when the patient is NPO. Note that serum electrolytes (Na, K, glucose, urea, creatinine) should be ordered prior to IV fluid administration as per the hospital’s Fluid and Electrolyte Administration in Children Clinical Practice Guideline. Request Haematology and Anesthesia consults.

2.2.2 Pre-operative Checklist

- Check patient’s electronic health record to identify if the Blood Transfusion Laboratory has blood group and red cell phenotype on file. If there is no phenotype in the patient’s electronic health record, call the Blood Transfusion Laboratory, extension 206208, to inform them that this is a sickle cell patient going to the O.R. If the patient is expected to need blood transfusion pre-op or intra-op, ensure there is a current sample for Type and Screen (indicate Sickle Cell Disease as diagnosis).

- Check that blood required and Sickle Cell Disease are indicated on the O.R. list.
2.2.3 Unless there are contraindications, initiation of incentive spirometry and referral to physiotherapy is indicated. Note that nursing may initiate incentive spirometry prior to assessment by physiotherapy. See Cardiopulmonary Physiotherapy policy.

2.2.4 The patient should be well oxygenated pre-operatively, with O₂ applied at 2 liters per minute when the patient is called to the operating room (oxygen should be applied for at least 15 minutes prior to surgery).

2.2.5 Keep patient warm with blankets.

### 2.3 Anesthetic Management

- It is important to prevent hypothermia as this can trigger sickling. Measures to ensure normothermia include pre-warming the operating room and/or use of forced-air warming (e.g., Bair Hugger).
- The patient should be well oxygenated pre-operatively; induction and intubation should be undertaken with little or no hypoxic insult. Monitor O₂ saturation closely.
- Avoid prolonged use of tourniquets.

### 2.4 Post-operative Management

2.4.1 Post-op care for patients with Sickle Cell Disease must emphasize pulmonary toilet and avoidance of hypoxia, hypotension, acidosis, and stasis. This begins in the operating room, with assurance that the patient does not become hypothermic, hypoxic, or hypotensive during emergence from anesthesia. Before extubation the patient should be well awake, ventilating and oxygenating well.

2.4.2 In the recovery ward, assess the patient before transferring the child to a paediatric ward. Request a chest x-ray, if there is any concern about respiratory function.

2.4.3 Patients should be monitored for O₂ saturation (aim to keep O₂ sat ≥95%); unless contraindicated, physiotherapy and incentive spirometry should be requested for all children.

2.4.4 Maintain hydration to prevent vasoconstriction, hypoperfusion, and microvascular occlusion, which ultimately lead to sickling. However, avoid over-hydration because pulmonary interstitial edema can lead to hypoxia and a sickling crisis.
2.4.5  Keep patient warm with blankets.

2.4.6  Provide appropriate analgesia so that the patient is co-operative with ambulation and pulmonary clearing and easily aroused from sleep.

3.0   References


Revised by (2016)

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Appendix A

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Peri-Operative Management: Guidelines for In-patient Management of Children with Sickle Cell Disease

Attachments:

Revision History.docx