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

Version: 4

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.[6] 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. [3]

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[6].

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[2] 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. However, regardless of clinical status, planned procedure and anesthetic, all patients with a diagnosis of Sickle Cell should have a pre-procedural consult with Haematology and the Pre-Anesthesia Clinic.

2.0 Sickle Cell Peri-operative Management Standard of Care

Sickle Cell Peri-operative Management Care Pathway

	PRE-OPERATIVE	PRE-OPERATIVE IN-PATIENT and INTRA-OPERATIVE	POST-OPERATIVE MANAGEMENT
GOALS	1. Admit all patients 1 day prior to surgery 2. Assess for risk factors and risk of potential complications 3. Child/family are advised of pre-op bath. Wipes to be used upon arrival. Refer to procedure document	1. Avoid hypothermia, hypoxia, and/or hypotension 2. Ensure child is awake, ventilating, and oxygenating well prior to extubating	1. Pulmonary toilet/no 2. Avoid hypoxia, hypotension, acidosis, and/or stasis 3. Keep child warm 4. Ensure adequate pain control

ASSESSMENT	• Complete thorough examination
	• Ensure that child does not have any acute illness
	• Postpone procedure if child is not healthy
	• *Consider transfusion for the following children:*
	  • undergoing major procedure i.e. thoracotomy, laparotomy, TJA;
	  • ill and surgery cannot be postponed;
	  • significant history of pulmonary disease, stroke, etc; and/or
	  • child with hemoglobin 15 g/L below baseline
	  • Less than 15g/L below baseline for urgent surgery
	  • Less than 85g/L as a baseline, discuss with Haematology 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 fellow/staff
		• Request Hematology and Anesthesia consults
		• **Complete pre-operative checklist:**
		  ✓ Ensure blood group and red cell phenotype are on patient's chart
		  ✓ If child 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 requisition for the issuing of blood products indicated 'Sickle Cell Disease' in addition to 'Pre-op'
		  ✓ If no contraindications, start incentive spirometer and refer to physiotherapy. See Cardiopulmonary Physiotherapy policy
		  ✓ Ensure child is well oxygenated pre-operatively; apply O_2 at 2L/min when patient is called to OR (initiate O_2 15 minutes prior to OR and continue O_2 on route to OR)
		  ✓ Ensure child is warm, apply warming blankets (from warming device)
		• **Anesthetic Management:**
		  ✓ Pre-warm OR and/or use forced air-warming i.e. Bair Hugger to ensure normothermia and prevent sickling due to hypothermia
		  ✓ Ensure child is well oxygenated pre-operatively
		  ✓ Induction and intubation should be undertaken with little or no hypoxic insult
		  ✓ Monitor O_2 closely
			• In the recovery ward, assessed thoroughly before transferring to in-patient unit
			• Request chest x-ray in the recovery room if there are concerns about respiratory dysfunction
			• Administer O_2 2L for 24 hours post op
			• Monitor O_2 saturation closely, ensure ≥95%
			• If no contraindications, start incentive spirometer and refer to physiotherapy. See Cardiopulmonary Physiotherapy policy

DIET & IV FLUIDS	• Review NPO requirement prior to surgery	• IV and PO fluids at maintenance • Maintain NPO status • When NPO administer IV fluids at maintenance • Refer to Fluid and Electrolyte Guidelines	• IV and PO fluids at maintenance • Use only warm or room temperature fluids post op • Maintain hydration to prevent vasoconstriction, hypo-perfusion, and microvascular occlusion (to prevent sickling) • Avoid over-hydration (to prevent pulmonary interstitial edema which can lead to hypoxia and sickling crisis)
LABS & MEDICATION		• Complete serum electrolytes (Na+, K+, glucose, urea, creatinine) prior to IV fluid administration	• Provide appropriate analgesia to ensure that child participates in ambulation and pulmonary clearing

PRINTABLE VERSION

3.0 References

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Attachments:

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