

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

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### 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.<sup>6</sup> 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.<sup>3</sup>

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<sup>6</sup>.

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<sup>2</sup> 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.

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## 2.0 Pathway

### Sickle Cell Peri-operative Management Care Pathway

	PRE-OPERATIVE	PRE-OPERATIVE INPATIENT and INTRA-OPERATIVE	POST-OPERATIVE MANAGEMENT
GOALS	<ol style="list-style-type: none"> <li>1. Admit all patients 1 day prior to surgery</li> <li>2. Assess for risk factors and risk of potential complications</li> <li>3. Child/family are advised of pre-op bath. Wipes to be used upon arrival. Refer to <a href="#">procedure document</a></li> </ol>	<ol style="list-style-type: none"> <li>1. Avoid hypothermia, hypoxia, and/or hypoxemia</li> <li>2. Ensure child is awake, ventilating, and oxygenating well prior to extubating</li> </ol>	<ol style="list-style-type: none"> <li>1. Pulmonary toilet/ing</li> <li>2. Avoid hypoxia, hypotension, acidosis, and/or stress</li> <li>3. Keep child warm</li> <li>4. Ensure adequate pain control</li> </ol>
ASSESSMENT	<ul style="list-style-type: none"> <li>• Complete thorough examination</li> <li>• Ensure that child does not have any acute illness</li> <li>• Postpone procedure if child is not healthy</li> <li>• Consider transfusion for the following children: <ul style="list-style-type: none"> <li>• undergoing major procedure i.e. thoracotomy, laparotomy, TJA;</li> <li>• ill and surgery cannot be postponed;</li> <li>• significant history of pulmonary disease, stroke, etc; and/or</li> <li>• child with hemoglobin 15 g/L below baseline</li> <li>• Less than 15g/L, below baseline for urgent surgery</li> <li>• Less than 85g/L, as a baseline, discuss with Hematology Team</li> </ul> </li> <li>• Plan general anesthetic carefully for elective procedures in collaboration with Sick Cell Team</li> <li>• Consider stroke risk and previous episodes of <b>Acute Chest Syndrome</b> as these can lead to post-op complications</li> <li>• Discuss any issues with Hematology fellow/attf</li> </ul>	<ul style="list-style-type: none"> <li>• Request Hematology and Anesthesia consults</li> <li>• Complete pre-operative checklist: <ul style="list-style-type: none"> <li>✓ Ensure blood group and red cell phenotype are on patient's chart</li> <li>✓ 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 Sick Cell Disease as diagnosis)</li> <li>✓ Ensure that blood required and Sick Cell Disease are indicated on OR list</li> <li>✓ If transfusion ordered pre-op, ensure the requisition for the issuing of blood products indicated 'Sickle Cell Disease' in addition to 'Pre-op'</li> <li>✓ If no contraindications, start incentive spirometer and refer to physiotherapy. See <a href="#">Cardiopulmonary Physiotherapy</a> policy</li> <li>✓ Ensure child is well oxygenated pre-operatively; apply O<sub>2</sub> at 2L/min when patient is called to OR (initiate O<sub>2</sub> 15 minutes prior to OR and continue O<sub>2</sub> on route to OR)</li> <li>✓ Ensure child is warm, apply warming blankets (from warming device)</li> </ul> </li> <li>• Anesthetic Management: <ul style="list-style-type: none"> <li>✓ Pre-warm OR and/or use forced air-warming i.e. Bair Hugger to ensure normothermia and prevent shivering due to hypothermia</li> <li>✓ Ensure child is well oxygenated pre-operatively</li> <li>✓ Induction and intubation should be undertaken with little or no hypoxic insult</li> <li>✓ Monitor O<sub>2</sub> closely</li> </ul> </li> </ul>	<ul style="list-style-type: none"> <li>• In the recovery ward, assesses thoroughly before transferring to in-patient unit</li> <li>• Request chest x-ray in the recovery room if there are concerns about respiratory dysfunction</li> <li>• Administer O<sub>2</sub> 2L for 24 hours post op</li> <li>• Monitor O<sub>2</sub> saturation closely, ensure 95%</li> <li>• If no contraindications, start incentive spirometer and refer to physiotherapy. See <a href="#">Cardiopulmonary Physiotherapy</a> policy</li> </ul>
DRUGS & IV FLUIDS	<ul style="list-style-type: none"> <li>• Review NPO requirement prior to surgery</li> </ul>	<ul style="list-style-type: none"> <li>• IV and PO fluids at maintenance</li> <li>• Maintain NPO status</li> <li>• When NPO administer IV fluids at maintenance</li> <li>• Refer to <a href="#">Fluid and Electrolyte Guidelines</a></li> </ul>	<ul style="list-style-type: none"> <li>• IV and PO fluids at maintenance</li> <li>• Use only warm or room temperature fluids post op</li> <li>• Maintain hydration to prevent vasoconstriction, hypo-perfusion, and microvascular occlusion (to prevent sickling)</li> <li>• Avoid over-hydration (to prevent pulmonary interstitial edema which can lead to hypoxia and sickling crisis)</li> </ul>
LABS & MONITORING		<ul style="list-style-type: none"> <li>• Complete serum electrolytes (Na<sup>+</sup>, K<sup>+</sup>, glucose, urea, creatinine) prior to IV fluid administration</li> </ul>	<ul style="list-style-type: none"> <li>• Provide econorboic analgesia to ensure that child participates in ambulation and pulmonary clearing</li> </ul>

## PRINTABLE VERSION

## 3.0 References

1. Platt OS, Brambilla DJ, Rosse WF, et al. Mortality in sickle cell disease: life expectancy and risk factors for early death. *N Engl J Med.* 1994;330:1639–44.

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2. Griffin TC, Buchanan GR. Elective surgery in children with sickle cell disease without preoperative blood transfusion. *J Pediatr Surg*. 1993;28(5):681–85.
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4. Haberkern CM, Neumayr LD, Orringer EP, et al. Cholecystectomy in sickle cell anemia patients: perioperative outcome of 364 cases from the National Preoperative Transfusion Study. *Blood*. 1997;89:1533–42.
5. Ware RE, Filston HC. Surgical management of children with hemoglobinopathies. *Surg Clin North Am*. 1992;72:1223–36.
6. Howard J, Malfroy M. et al. The Transfusion Alternatives Preoperatively in Sickle Cell Disease (TAPS) study: a randomised, controlled, multicentre clinical trial. *Lancet*. 2013;381: 930-938.

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## Attachments:

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