Scope: Hospital-wide Patient Care
Document Type: Clinical Practice Guideline
Approved on: 2021-07-20
Next Review Date: 2023-07-20

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

Version: 3

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.

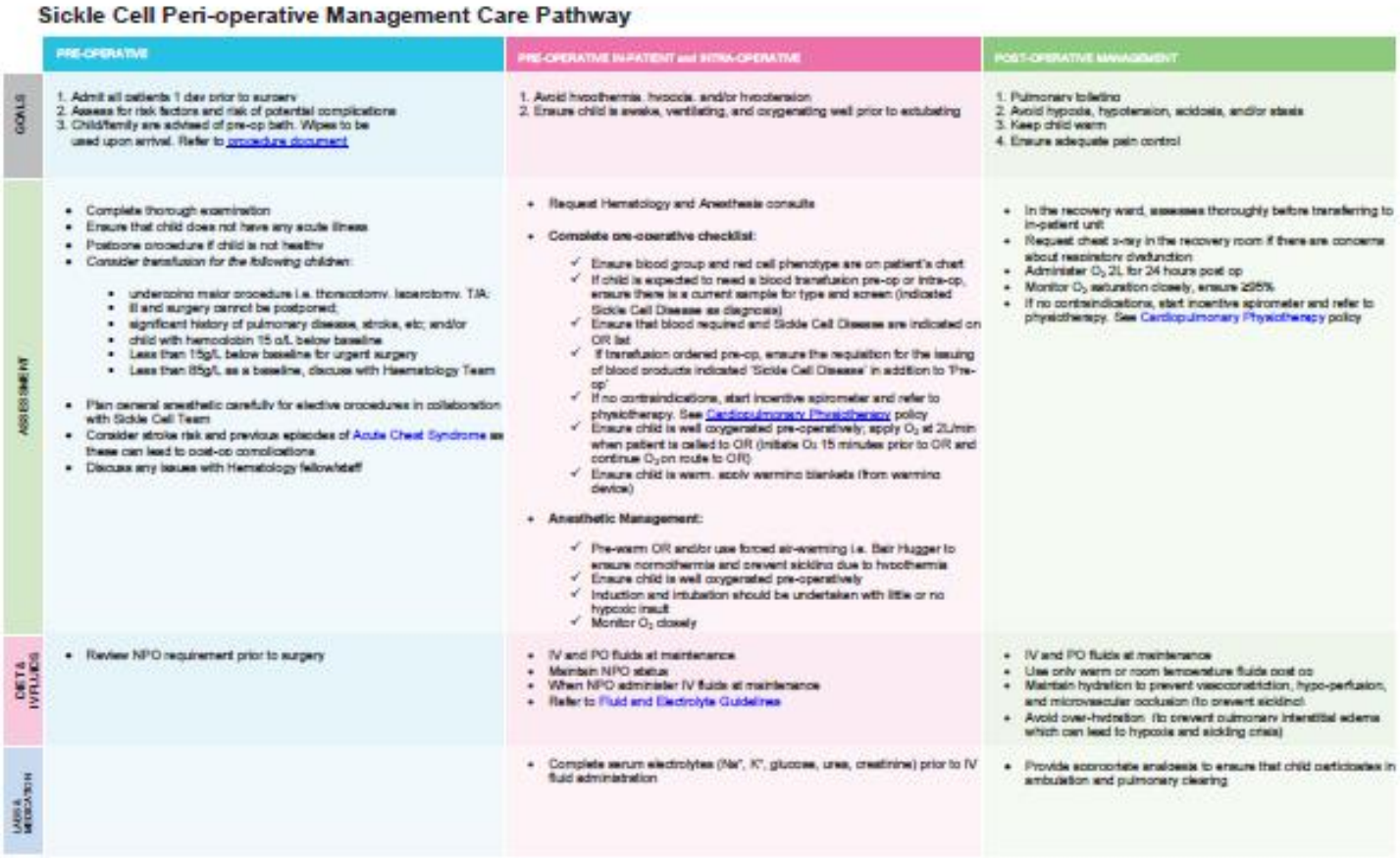
It has been shown that the correction of anemia and reduction in the percentage of haemoglobin S will prevent intra-operative and post-operative morbidity and mortality in sickle cell patients.[6] Historically, uncertainty has existed as to the benefits of simple pre-operative transfusion, given the 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.

2.0 Pathway



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.

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.

3. Vichinsky EP, Haberkern CM, Neumayr LD, et al. A comparison of conservative and aggressive transfusion regimens in the perioperative management of sickle cell disease. *N Engl J Med*. 1995;333(4):206–13.

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.

Reviewers

1. Carolyn Beck, MD, Staff Paediatrician, Division of Paediatric Medicine
2. Lynn Mack, RN, Quality Analyst 7BCD5.
3. Marcia Palmer, RN, Division of Haematology/Oncology
4. Melanie Kirby, MD, Staff Physician, Division of Haematology/Oncology
5. Melina Cheong, RN, Nurse Practitioner, Division of Haematology/Oncology
6. Marina Strzelecki, Pharmacist

Attachments:

Revision History.docx

SCD Periop 2021 FINAL.pdf