1.0 Background
By 4–5 months of age, splenomegaly develops in some infants with sickle cell disease, and by 12 months of age a palpable spleen is noted in nearly half. Although enlarged, the spleen does not properly perform its filtration function. However, its reservoir function is overactive: sequestration of large quantities of blood (often half or more of a child’s blood volume) can occur rapidly. This complication, termed acute splenic sequestration, is characterized by pooling of large quantities of sickled RBCs in the splenic red pulp, sudden enlargement of the spleen (within hours), and a precipitous decline in haemoglobin (Hb) and platelets, and an increase in reticulocytes.

Presentation is often (60%) associated with episodes of fever, suggesting an underlying viral etiology. Most commonly occurs in infants and young children between 6 months and 5 years of age with sickle cell anaemia. It may also occur in older patients with any sickle cell phenotype with or without chronic splenomegaly. Often there is no obvious triggering event.

2.0 Clinical/Laboratory Features
A child with an acute splenic sequestration presents with symptoms of:
- acute anaemia (pallor, tachycardia, frank cardiovascular collapse);
- splenomegaly/abdominal pain (pain in the left upper quadrant); and
- evidence of an active bone marrow response (increased reticulocytes) plus or minus thrombocytopenia.

Retrospective reviews have shown a first-episode mortality of as high as 14%. On physical examination, patients show signs of anaemia, hypovolemia, and an enlarged spleen (larger than baseline), sometimes massively so. Mild cases may be asymptomatic.

Haemoglobin concentration is at least 20g/L below the baseline steady state. In severe cases, haemoglobin may decline to life-threatening levels. Reticulocyte counts are usually elevated, which distinguishes this condition from aplastic crisis. The platelet count often declines to <50 X 10^9/L.

The mainstay of management is transfusion to provide circulating erythrocytes and volume. Risk of recurrence is approximately 40–50%, usually within 3 years. Because it is not possible to predict which children will have recurrent attacks, most experts recommend splenectomy after the first major attack (for patients >2 years old), or chronic transfusion to maintain a haemoglobin S level under 50% until the patient can get to surgery once all relevant immunizations have been completed.

3.0 Clinical Practice Recommendations

Statement of Evidence: Recommendations were made by expert group consensus. (Grade C)
### Acute Splenic Sequestration: Guidelines for Management in Children with Sickle Cell Disease

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#### 3.1 Emergency Department Treatment

3.1.1 Conduct a history and physical exam with emphasis on signs and symptoms of cardiovascular collapse (shock), anemia, and hypovolemia. If the spleen is palpable, attempt to verify with the parent/clinic chart previous spleen size for comparison.

3.1.2 Tests: CBC, differential, reticulocyte count, blood type and cross-match, O₂ saturation and Arterial Blood Gas (ABG). Establish IV access noting 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 Clinical Practice Guideline.

3.1.3 Patients with Acute Splenic Sequestration should be admitted to the Paediatric Medicine Unit (7BCDE), or to the CCU if unstable, following a discussion with the Haematology consult fellow (or the Haematology fellow on-call after hours), who should notify the Sickle Cell Team by telephone (416-813-6443).

3.1.4 If hemoglobin is greater than 20g/L below baseline, transfuse with appropriately cross matched Packed Red Blood Cells (PRBC) not exceeding 100g/L. If hemodynamically unstable, give immediate IV fluid bolus followed by PRBC and call the Haematology team.

#### 3.2 In-patient Management

3.2.1 While the child is an in-patient, take vitals as per BedsidePEWS, careful repeated physical assessments (q4–6h) for spleen size (measure with tape and record), and Hb measurements (q8-12h). The patient’s heart and respiratory rates and O₂ saturation should be recorded on a monitor. Patients should be on a cardiac or O₂ monitor.

3.2.2 If hemoglobin is greater than 20g/L below baseline, transfuse as soon as possible with appropriately cross matched PRBC not exceeding 100g/L and notify the Haematology team. Use available phenotypically matched blood. If hemodynamically unstable, give immediate IV fluid bolus followed by PRBC and follow BedsidePEWS recommendations.

3.2.3 In hospital, continue regularly scheduled medications.

3.2.4 Administer oxygen to keep O₂ saturation ≥95%.

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3.2.5 An acute sequestration episode usually resolves within 2–5 days. When there is evidence of rising haemoglobin and diminishing spleen size, the patient can be discharged, with close out-patient follow-up by the Sickle Cell Team.

3.2.6 For weeks to months following an episode, some patients have persistent splenomegaly and hypersplenism, with lower-than-usual Hb and platelet values. Blood count should be monitored.

3.2.7 Decisions regarding splenectomy will be made in follow-up with the Sickle Cell Team as an outpatient. Prior to discharge, ensure that a follow-up appointment with the Sickle Cell Team has been confirmed (appointment should occur within two weeks of discharge).

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4.0 References


Attachments:

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