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 10⁹/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 Guideline

### Acute Splenic Sequestration: Guidelines for Management in Children with Sickle Cell Disease

**ED Management**
- Gather history and complete physical exam
- **Complete tests:**
  - Establish IV access
  - Ensure child is on cardiopulmonary monitoring and O₂
  - Hemoglobin (Hb) and reticulocyte count
  - Transfusion volume
  - Hematocrit (Hct) following transfusion
- **Inpatient Management:**
  - Child must be on cardiac or O₂ monitor
  - Monitor vital signs as per BedsidePEWs
  - Repeat physical assessment:
    - Spleen size: q4-6h (measure with tape and record)
    - Hct q12h
  - Hemoglobin (Hb) and reticulocyte count: baseline, transfusion as soon as possible with cross matched PRBC not exceeding 10g/dL, and notify Haematology Team; Transfusion volume: 5-10mL/kg, determined in consultation with Haematology Team, use phenotypically matched blood. If unstable, give IV fluid bolus followed by PRBC
  - Refer to Haematology (notify Sickle Cell Team) or keep SpO₂ >95%

**Inpatient Management**
- Child discharged home from inpatient unit with appropriate follow-up if:
  - Evidence of rising haemoglobin and diminishing spleen size
  - Hemoglobin (Hb) q6-12h
  - Refer to Admission Planning Process document
  - Follow regularly scheduled medications
  - Monitor O₂ and keep SpO₂ >95%

**ED Management**
- Child presents in ED with symptoms of Acute Splenic Sequestration
- Gather history and complete physical exam

**Symptoms of Acute Splenic Sequestration:**
- Acute upper abdominal pain (in the left upper quadrant), and parascapular pain
- Evidence of an splenomegaly (enlarged spleen size) on abdominal ultrasound
- Headache
- Abdominal distension
- Hypovolemic shock

**Inpatient Management**
- Child must be on cardiac or O₂ monitor
- Monitor vital signs as per BedsidePEWs
- Repeat physical assessment:
  - Spleen size: q4-6h (measure with tape and record)
  - Hct q12h
- Hemoglobin (Hb) and reticulocyte count: baseline, transfusion as soon as possible with cross matched PRBC not exceeding 10g/dL, and notify Haematology Team; Transfusion volume: 5-10mL/kg, determined in consultation with Haematology Team, use phenotypically matched blood. If unstable, give IV fluid bolus followed by PRBC and follow BedsidePEWs recommendations
- Refer to Haematology (notify Sickle Cell Team) or keep SpO₂ >95%

**Transfer to CCU**
- Refer to Haematology (notify Sickle Cell Team) or keep SpO₂ >95%

**Admit to Pediatric Medicine**
- Refer to Haematology (notify Sickle Cell Team) or keep SpO₂ >95%

**Follow up**
- Confirmed within 2 weeks of discharge
- Evidence of rising haemoglobin and diminishing spleen size
- Hemoglobin (Hb) q6-12h
- Refer to Admission Planning Process document
- Follow regularly scheduled medications
- Monitor O₂ and keep SpO₂ >95%

**Child discharged home from inpatient unit with appropriate follow-up if:**
- Evidence of rising haemoglobin and diminishing spleen size
- Hemoglobin (Hb) q6-12h
- Refer to Admission Planning Process document
- Follow regularly scheduled medications
- Monitor O₂ and keep SpO₂ >95%

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### References


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Attachments:

- Revision History.docx
- SC_Clinic Follow Up Revised 2021_FINAL.pdf
- Splenic Sequestration Care Pathway 2021 Final.pdf