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

Child presents in ED with symptoms of Acute Splenic Sequestration:
- Acute internal pain, vomiting, pain in the left upper quadrant, and evidence of an acute bone marrow response (increased reticulocytes, plus or minus thrombocytopenia).
- Skin rash, abdominal pain, plus or minus thrombocytopenia.
- Disappearance of splenic size on abdominal x-ray.

ED Management:
- Complete tests:
  - Initiate Sickle Cell Fever order set in Epic as indicated
  - Bolus followed by PRBC
  - Phenotypically matched blood
- Consultation with Haematology Team
- Transfusion volume not exceeding 10% of baseline, transfuse as soon as possible with cross matched PRBC not exceeding 10% of baseline, and notify Haematology Team.
- Transfusion volume ≥ 10%, determined in consultation with Haematology Team, use phenotypically matched blood. If unstable, give fluid bolus followed by PRBC.
- Initiate Sickle Cell Fever order set in Epic as indicated
- Consult Haematology (notify Sickle Cell Team)

Is the child stable?
- YES
  - Transfer to CCU
  - Consult Haematology (notify Sickle Cell Team)
- NO
  - NO
  - Admit to Pediatric Medicine

Inpatient Management:
- Child must be on cardiac or O2 monitor
- Continuous renal monitoring
- Monitor vital signs as per Best Practice
- Repeated physical assessment:
  - Splenic size q4-6h (measure with tape and record)
  - Labs q12h
- Hemoglobin ≤ 8 g/dl, below baseline, transfuse as soon as possible with cross matched PRBC not exceeding 10% of baseline, and notify Haematology team. Transfusion volume ≥10%, determined in consultation with Haematology Team, use phenotypically matched blood. If unstable, give fluid bolus followed by PRBC and follow Best Practice recommendations
- Continue regularly scheduled medications
- Administer O2 to keep SpO2 ≥ 95%

Child discharged home from inpatient unit with appropriate follow-up:
- Evidence of rising hemoglobin and diminishing splenic size
- Reference to discharge planning process
- Child has stable vital signs i.e. stable unless clear viral source
- Fruitful feeds and voiding by mouth
- Child’s pain is controlled by oral analgesics
- Extreme risk for readmission (high risk: ≥ 3 admissions for VOC in the last 12 months)
- Admission at high risk for readmission must be referred for CCAC for follow up and seen in clinic setting
- 1 week of discharge
- Does not have respiratory distress
- Follow up confirmed within 2 weeks of discharge (including blood culture follow-up)

**REFERENCES**


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**Attachments:**

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