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 Guideline

Child presents in ED with symptoms of Acute Splenic Sequestration

- Complete tests
- Establish IV access
- Ensure child is on continuous intravenous and O2
- If hematocrit is >26%, below baseline, transfuse as soon as possible with cross-matched PRBC not exceeding 10U, and notify Hematology Team; Transfusion volume 3-5L. Determine need for consultation with Hematology Team. Use phenotypically matched blood. If unstable, give IV fluid bolus followed by PRBC.
- Initiate Sickle Cell Fever order set in Epi as indicated

Gather history and complete physical exam

ED Management

- Blood work: CBC, reticulocyte count, blood typing and cross-match, and serum Na, K, glucose, creatinine
- Os sats and arterial blood gases
- Emphasis on signs of cardiovascular collapse (shelf), aura, and hypovolemia
- Evidence of an active bone marrow response (increased reticulocytes, platelet count)

Symptoms of Acute Splenic Sequestration:
- Acute pain in the left upper quadrant, and evidence of an active bone marrow response (increased reticulocytes, platelet count)
- Sudden hypotension, shock, abnormal cardiopulmonary monitoring
- Transfusion volume 3-5L. Determine need for consultation with Hematology Team. Use phenotypically matched blood. If unstable, give IV fluid bolus followed by PRBC.

Consult Hematology (notify Sickle Cell Team)

Inpatient Management

- Child must be on cardiac or O2 monitor
- Monitor vital signs as per BedsidePWEs
- Repeat physical assessment:
  - Spleen size q6h (measure with tape and record)
  - II at q12h
- If hematocrit is >26%, below baseline, transfuse as soon as possible with cross-matched PRBC not exceeding 10U, and notify Hematology Team; Transfusion volume 3-5L. Determine need for consultation with Hematology Team. Use phenotypically matched blood. If unstable, give IV fluid bolus followed by PRBC and follow BedsidePWEs recommendations
- Continue regularly scheduled medications
- Administer O2 to keep SpO2 ≥ 95%

Is the child stable?

NO

- Transfer to CCU
- Admit to Pediatric Medicine

YES

Child discharged home from inpatient unit with appropriate follow-up:

- Evidence of rising hematocrit and diminishing spleen size
- Refer to discharge planning process document
- Child has stable vital signs i.e. stable unless clear vital source;
- Tending fluids and medications by mouth
- Child’s pain is controlled by oral analgesics;
- Cautiously wean IV fluids and wean from any high risk: all admissions for VOC or IV adms for ACS in the last 12 months
- Admissions at high risk for readmission must be referred for COLAC for follow-up and may be seen in clinic setting 14 days of discharge
- Does not have respiratory distress; and
- Follow-up confirmed with in 2 weeks of discharge (including blood culture follow-up)

4.0 References


**Attachments:**

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