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

**Acute Splenic Sequestration**

**Guideline**

- Acute splenic sequestration is a life-threatening complication of sickle cell disease characterized by a sudden dilation of the splenic pulp.

**Symptoms of Acute Splenic Sequestration**

- Sudden onset of abdominal pain, usually left upper quadrant.
- Evidence of an elevated white blood cell count, increased reticulocyte count, and decreased hemoglobin.
- Hypotension, tachycardia, and pallor are indicative of frank cardiovascular collapse.

**ED Management**

1. Gather history and complete physical exam.
2. Complete tests:
   - Establish IV access
   - Ensure child is on capillarymonitoring and O2
   - Echocardiogram: at end of baseline, transfuse as soon as possible with cross matched PRBC
   - Transfusion volume: 2.5 g/kg, determined in consultation with Haematology Team. Use phenotypicallymatched blood. If unavailable, give fixed blood followed by PRBC.
3. Consult Haematology (notify Sickle Cell Team).
4. Establish Sickle Cell Fever order in Epic as indicated.
5. Child presents in ED with symptoms of Acute Splenic Sequestration:
   - Child must be on cardiac or O2 monitor
   - Monitor vital signs as per BedsidePEWs
   - Repeat physical assessment:
     - Spleen size: q4h - measure with tape and record
     - O2 sat q12h
   - Echocardiogram: at end of baseline, transfuse as soon as possible with cross matched PRBC not exceeding 10 g/kg.
   - Notify Hematology Team: Tranfusion volume 2.5 g/kg, determined in consultation with Haematology Team. Use phenotypicallymatched blood. If unavailable, give fixed blood followed by PRBC and follow BedsidePEWs recommendations.
   - Continue regularly scheduled medications.
   - Administer O2 to keep SpO2 ≥95%

**Inpatient Management**

- Child discharged home from inpatient unit with appropriate follow-up:
  - Evidence of rising hemoglobin and diminishing spleen size
  - Refer to discharge planning process document
  - Child has stable vital signs i.e. stable unless clear viral source
  - Tend to fluids and medications by mouth
  - Child's pain is controlled by oral analgesics
  - Continue risk for readmission: high risk: admission for VOD or IV1 admission for ACS in the last 12 months
  - Discharge at high risk for readmission must be referred for CCAC for follow up and be seen in clinic setting 1-2 days of discharge
  - Does not have respiratory distress
  - Follow up continued within 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