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

Sickle Cell Anemia: Splenic Sequestration Management Guidelines

Child presents in ED with symptoms of Acute Splenic Sequestration

Gather history and complete physical exam

ED Management
- Complete tests
- Establish IV access
- Ensure child is on cardiopulmonary monitoring and O₂
data
- If hemoglobin is ≤28 g/L, below baseline, transfuse as soon as possible with cross-matched PRBC, not exceeding 100 ml/kg transfusion volume
- Transfusing volume ≤500 ml
- If unstable, consult with Haematology Team
- Use phenotypically matched blood. If unavailable, give IV fluid bolus followed by PRBC
- Initiate Sickle Cell Fever order set in Epic as indicated

Transfer to CCU

Admit to Pediatric Medicine

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)
  - labs q12h
- If hemoglobin is ≤28 g/L, below baseline, transfuse as soon as possible with cross-matched PRBC, not exceeding 100 ml/kg, and notify Haematology Team
- Transfusing volume ≤150 ml
- If unstable and if unable to unilaterally give IV fluid bolus followed by PRBC and follow BedsidePEWs recommendations
  - Continue regularly scheduled medications
  - Administer O₂ to keep SpO₂ ≥95%

Child discharged home from inpatient unit with appropriate follow-up if:
- Evidence of rising hemoglobin and diminishing spleen size
- Pain is controlled by oral analgesics
- No signs of hypovolemia
- Child has stable vital signs i.e. stable unless clear vital source
- Expand fluids and medications by mouth
- Child pain is controlled by oral analgesics
- Extensive risk for readmission (high risk: ≥3 admissions for VOC or ≥1 admission for ACS in the last 12 months)
- Hospitalization at time of 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; and
- Follow-up confirmed within 3 weeks of discharge (including blood culture follow-up)

Symptoms of Acute Splenic Sequestration:
- Acute pain (abdominal pain in the left upper quadrant), and evidence of an acute bone marrow response (increased reticulocyte count or increased thrombocytopenia)
- Palpable splenomegaly (spleen palpable in the left upper quadrant)
- Hypovolemia (systolic BP<100 mmHg, pulse>100 bpm)
- Evidence of cardiovascular collapse (shock)

Gather history and complete physical exam

NO

YES

Is the child stable?

Transfer to CCU

Admit to Pediatric Medicine

3.0 Clinical Practice Guideline

4.0 References


Attachments:

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