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

Symptoms of Acute Splenic Sequestration:
- Acute onset of abdominal pain in the left upper quadrant and evidence of an absent or hyporesponsive (increased reticulocyte count) peripheral blood smear.
- Painless splenomegaly.
- Tachycardia.
- Hypotension.
- Evidence of frank cardiovascular collapse.
- Anemia.
- Pallor.
- Tachypnea.
- Hypovolemia.
- Evidence of an active bone marrow response (increased reticulocyte count).
- Evidence of shock.

ED Management
- Complete the history and physical exam.
- Establish IV access.
- Initiate Sickle Cell Fever order set in Epic as indicated.
- Administer O2, fluids, and medications as per BedsidePEWs.
- Monitor vital signs.
- Perform physical assessment.
- Obtain blood work (CBC, reticulocyte count, platelet count, prothrombin time, partial thromboplastin time, electrolytes, glucose, creatinine, urinalysis, blood cultures).
- Obtain imaging studies as indicated (ultrasound, CT scan).
- Consult Haematology (notify Sickle Cell Team).

Inpatient Management
- Child must be on cardiac or O2 monitor.
- Monitor vital signs as per BedsidePEWs.
- Obtain repeat physical assessment:
  - Spleen size q4h (measure with tape and record).
  - Lab q8h.
- Obtain blood work:
  - CBC, reticulocyte count, platelet count, prothrombin time, partial thromboplastin time, electrolytes, glucose, creatinine, urinalysis, blood cultures.
  - Imaging studies as indicated (ultrasound, CT scan).
- Consult Haematology.
- Order crossmatched PRBCs if hemoglobin is <8 g/dL.
- Transfuse as soon as possible with crossmatched PRBCs.
- Continue regularly scheduled medications.
- Discharge the child from the inpatient unit with appropriate follow-up:
  - Evidence of an active bone marrow response and diminishing spleen size.
  - Refer to discharge planning process (document). 
  - Child to have stable vital signs (i.e., stable unless clear viral source).
  - Tending fluids and medications by mouth.
  - Child pain is controlled by oral analgesia.
  - Evidence of repeat admission: high-risk, readmission for VOC or inpatient for ACS in the last 12 months.
  - Admissions at high risk for readmission must be referred for OAC for follow-up and seen in clinic setting 14 days of discharge.
  - Does not have respiratory distress and follows up confirmed within 2 weeks of discharge (including blood culture follow-up).

Child discharged home from inpatient unit with appropriate follow-up:
- Evidence of an active bone marrow response and diminishing spleen size.
- Refer to discharge planning process (document).
- Child to have stable vital signs (i.e., stable unless clear viral source).
- Tending fluids and medications by mouth.
- Child pain is controlled by oral analgesia.
- Evidence of repeat admission: high-risk, readmission for VOC or inpatient for ACS in the last 12 months.
- Admissions at high risk for readmission must be referred for OAC for follow-up and seen in clinic setting 14 days of discharge.
- Does not have respiratory distress, and
- Follow-up confirmed 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