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.

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.
Acute Splenic Sequestration: Guidelines for Management in Children with Sickle Cell Disease

**Inpatient Management**
- Child must be on cardiac or \( \mathrm{O}_2 \) monitor
- Monitor vital signs as per BedsidePEWs
- Repeat physical assessment:
  - Spleen size: q4-h (measured with tape and recorded)
  - \( \mathrm{Fi}O_2\)-q12-h
- If hemoglobin is ≥20g/L below baseline, transfuse as soon as possible with cross matched PRBC not exceeding 100g/L and notify Haematology team. Use phenotypically matched blood. If unstable, give IV fluid bolus followed by PRBC.
- Initiate Sickle Cell Fever order set in Epic as indicated

**ED Management**
- Complete tests
- Establish IV access
- Ensure child is on cardiopulmonary monitoring and \( \mathrm{O}_2 \) ≥95%
- If hemoglobin is ≥20g/L below baseline, transfuse as soon as possible with cross matched PRBC not exceeding 100g/L and notify Haematology team. Use phenotypically matched blood. If unstable, give IV fluid bolus followed by PRBC.
- Initiate Sickle Cell Fever order set in Epic as indicated

**Symptoms of Acute Splenic Sequestration:**
- Acute anemia (pallor, tachycardia, tachypnea, cardiovascular collapse);
- Evidence of an active bone marrow response (increased reticulocytes) plus or minus thrombocytopenia
- Splenomegaly
- Evidence of rising hemoglobin and diminishing spleen size
- Acute anemia (pallor, tachycardia, tachypnea, cardiovascular collapse);
- Evidence of an active bone marrow response (increased reticulocytes) plus or minus thrombocytopenia

**Blood work:** CBC, reticulocyte count, blood typing and cross match, and serum Na, K, glucose, creatinine
- \( \mathrm{O}_2 \) saturation and arterial blood gases
- Urine output
- ECG

**Gather history and complete physical exam**
- Child presents in ED with symptoms of Acute Splenic Sequestration
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  - Continue regularly scheduled medications
  - Admit child to keep \( \mathrm{SpO}_2 \) ≥95%

**Child discharged home from inpatient unit with appropriate follow-up:**
- Evidence of rising haemoglobin and diminishing spleen size;
- Blood count (hb and platelet) should be monitored due to persistent splenomegaly and hypoplasentism;
- Refer to discharge planning process document;
- Child has stable vital signs i.e. stable unless clear viral source;
- Tolerating fluids and medications by mouth;
- Child’s pain is controlled by oral analgesics;
- Determine risk for readmission (high risk: ≥3 admissions for VOC or ≥1 admission for ACS in the last 12 months);
- All patients at high risk for readmission must be referred to CCAC for follow-up and be seen in clinic within 2 weeks of discharge;
- Does not have respiratory distress; and
- Follow up confirmed within 2 weeks of discharge (including blood culture follow-up).

**Document Scope:** Hospital-wide Patient Care

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References


Attachments:

- SC_clinic follow up.pdf
- Revision History.docx
- pathway_splenic_final.pdf

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