All preprinted orders and clinical care pathways for the management of sickle cell disease can be found on the APPHON website at:
https://www.apphon-rohppa.com/en/guidelines/sickle-cell-asplenia-guidelines

Initial Management of Sickle Cell Disease (SCD) – ED Pathway

Triage and Assessment

A child with SCD presenting to ED with fever or pain should be assessed within 30 minutes of arrival.

Check for signs of complications,

- Vaso-occlusive crisis (painful crisis)
- Fever – sepsis
- Acute chest syndrome
- Stroke
- Priapism
- Aplastic crisis
- Acute splenic sequestration.

General Sickle Cell Disease Management

1. **Pain**: Start analgesics promptly – treat pain aggressively.
   - Mild: acetaminophen, ibuprofen.
   - Moderate: Consider oral morphine or hydromorphone.
   - Moderate to severe: morphine or hydromorphone IV, repeat as needed, may need continuous infusion *https://www.apphon-rohppa.com/en/guidelines/sickle-cell-asplenia-guidelines*
2. **Fluids**:
   - Encourage oral fluids. May require IV fluid bolus 10 – 20 mL/kg
   - Avoid excess fluids to reduce risk of chest crisis.
   - Do **not** delay starting IV fluids or analgesics for topical anaesthetic cream. If unable to obtain IV access after 2 attempts, consider ultrasound guided IV placement or page anesthesia/anesthesia assistant.
3. **Antibiotics:** If patient presents with fever start IV Ceftriaxone *https://www.apphon-rohppa.com/en/guidelines/sickle-cell-asplenia-guidelines*
4. **Consultations:** May require blood transfusion – discuss with Hematology.

Contact Hematologist on call

Investigations:

- CBCD including reticulocyte count
- Blood group & cross match
- CRP, blood and urine cultures if febrile
- Serum Creatinine, BUN and electrolytes and LFTs if dehydrated or jaundice
- Chest X-ray if respiratory symptoms
- Other imaging as required

Vaso-occlusive crisis (painful crisis)	Fever – sepsis	Acute chest syndrome	Stroke	Acute splenic sequestration	Aplastic crisis	Priapism
Precipitated by dehydration, hypoxia or infection. All episodes of pain should be treated initially as vaso-occlusive disease as per Sickle VOC clinical pathway on the APPHON website. Chest pain may indicate an acute chest syndrome rather than as a vaso-occlusive episode if associated with respiratory symptoms. *Refer to clinical care pathway on the APPHON website.*	Patients are functionally asplenic and at greater risk for invasive disease by encapsulated organisms. **Specific management:** • Start IV Ceftriaxone as per the *ED SCD fever orders on APPHON website.* • Consider coverage for atypical organisms (Clarithromycin) if significant respiratory component. • Obtain appropriate cultures. ○ Blood, sputum, urine. If pain is also present, treat as vaso-occlusive crisis If cough or dyspnoea is present look and treat for acute chest syndrome.	Life threatening condition. Suspect if respiratory distress, hypoxia or chest pain. **Specific management:** • Oxygen to keep oxygen saturations > 93% or for comfort. • Analgesia as above • Start IV antibiotics – Ceftriaxone and Clarithromycin as per the *ED SCD fever orders on the APPHON website.* • Chest X-ray – but don't delay treatment. • Consider simple transfusion in consultation with hematology. Do NOT exceed a post transfusion Hb of 100g/L. • Early referral to PICU for respiratory support if significant hypoxia or respiratory distress. *Refer to clinical care pathway on the APPHON website.*	Can occur suddenly or as a complication of acute chest syndrome or aplastic crisis. **Specific management:** Neuroimaging required to determine if hemorrhagic or ischemic stroke. • MRI is modality of choice (ED or hematology to order). If not available, • CT - NO CONTRAST (risk of hyperviscosity). **Transfusion support:** • Options include initial simple transfusion to Hb 100 g/L followed by red cell exchange. *Refer to clinical care pathway on the APPHON website.*	Anemia (↓Hb >20g/L) with thrombocytopenia and acute splenomegaly. May present acutely shocked. **Specific management**: • Fluid resuscitation – NS 0.9% 10 -20 mL/kg Initial transfusion to aim for Hb of 50 - 60 g/L initially to ameliorate hemodynamic instability. • Do not increase Hb by > 30 g/L of presenting Hb with initial transfusion and do NOT exceed a post transfusion Hb of 100 g/L. Auto-transfusion may occur if hemoglobin is increased excessively or too quickly. This increases risk of stroke due to hyperviscosity. IV antibiotics if febrile as per the *ED SCD fever orders on the APPHON website.* *Refer to clinical care pathway on the APPHON website.*	An acute illness with decreased hemoglobin without a reticulocyte response (usually <1%). Usually associated with acute infection including parvovirus. Present with pallor +/- shock. **Specific management:** • Intravenous fluids and oral intake to a total of maintenance. • Transfuse red blood cells if patient is asymptomatic with anemia or Hb • <50 g/L (do NOT increase Hb by > 30 g/L of presenting Hb with initial transfusion). • Start IV antibiotics if febrile – Ceftriaxone as per the *ED SCD fever orders on the APPHON website.*	Two forms – intermittent or prolonged. **Specific management**: Do not use ice. • Simple measures e.g. moderate exercise, take a bath or shower. • Empty bladder – may need catheter • Analgesia, oxygen, hydration with alkalization of the urine should be commenced as soon as possible. Consult Pediatric Urologist and on-call hematologist.

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