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 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 (refer to IWK DDG for intermittent IV dosing), may need continuous infusion. Refer to Sickle Cell Disease Pain Admission Orders on APPHON website.
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. Refer to guidelines on APPHON website.
4. **Consultations:** May require blood transfusion – discuss with Hematology. Early PICU review and respiratory support if concern for acute chest crisis.

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 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 > 94% or for comfort. • Analgesia as above. • Start IV antibiotics – Ceftriaxone and Clarithromycin as per the ED SCD fever orders on 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 ED SCD fever orders on 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 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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