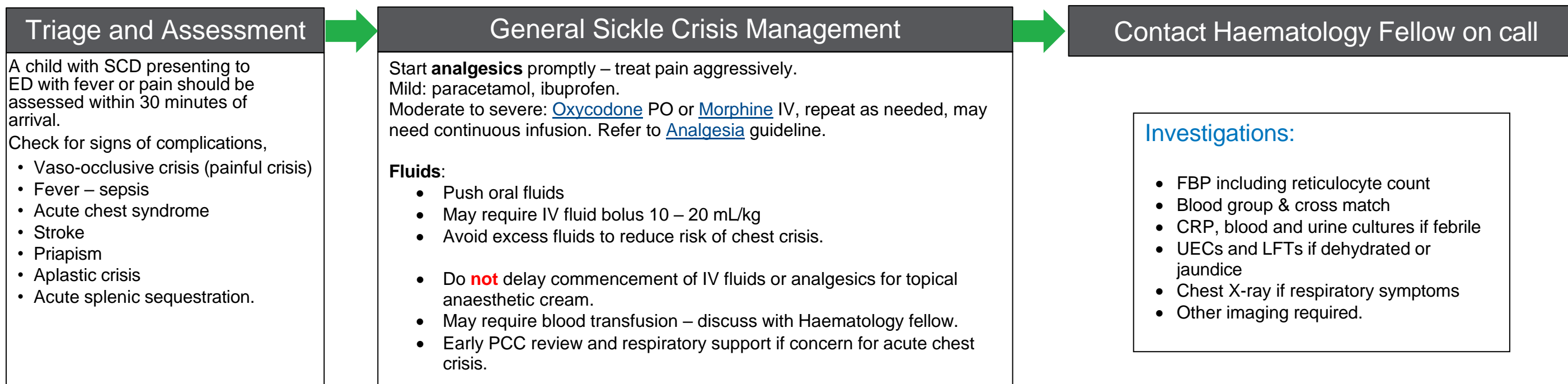


Initial Management of Sickle Cell Disease (SCD) – ED Pathway



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 Crisis Management

Start **analgesics** promptly – treat pain aggressively.

Mild: paracetamol, ibuprofen.
Moderate to severe: [Oxycodone](#) PO or [Morphine](#) IV, repeat as needed, may need continuous infusion. Refer to [Analgesia](#) guideline.

Fluids:

- Push oral fluids
- May require IV fluid bolus 10 – 20 mL/kg
- Avoid excess fluids to reduce risk of chest crisis.
- Do **not** delay commencement of IV fluids or analgesics for topical anaesthetic cream.
- May require blood transfusion – discuss with Haematology fellow.
- Early PCC review and respiratory support if concern for acute chest crisis.

Contact Haematology Fellow on call

Investigations:

- FBP including reticulocyte count
- Blood group & cross match
- CRP, blood and urine cultures if febrile
- UECs and LFTs if dehydrated or jaundice
- Chest X-ray if respiratory symptoms
- Other imaging required.

Vaso-occlusive crisis (painful crisis)	Fever – sepsis	Acute chest syndrome	Stroke	Priapism	Aplastic crisis	Acute splenic sequestration
<p>Precipitated by dehydration, hypoxia or infection.</p> <p>All episodes of pain should be treated initially as vaso-occlusive disease as per General Sickle Crisis Management.</p> <p>Chest pain may indicate an acute chest syndrome rather than as a vaso-occlusive episode if associated with respiratory symptoms.</p>	<p>Patients are functionally asplenic and at greater risk for invasive disease by encapsulated organisms.</p> <p>Specific management:</p> <ul style="list-style-type: none"> • Commence IV Ceftriaxone • Consider cover for atypical organisms (Azithromycin) if significant respiratory component • Obtain appropriate cultures <ul style="list-style-type: none"> ○ Blood, sputum, urine <p>If pain is also present, treat as vaso-occlusive crisis. If cough or dyspnoea is present look and treat for acute chest syndrome.</p>	<p>Life threatening condition. Suspect if respiratory distress, hypoxia or chest pain.</p> <p>Specific management:</p> <ul style="list-style-type: none"> • Oxygen to keep oxygen saturations > 96% or for comfort. • Analgesia • Commence IV antibiotics – Ceftriaxone and Azithromycin. • Chest X-ray – but this should not delay commencement of treatment. • Early referral to PCC for respiratory support if significant hypoxia or respiratory distress. 	<p>Can occur suddenly or as a complication of acute chest syndrome or aplastic crisis.</p> <p>Specific management:</p> <p>Neuroimaging required to determine if haemorrhagic or ischaemic stroke.</p> <ul style="list-style-type: none"> • MRI is modality of choice. <p>If not available,</p> <ul style="list-style-type: none"> • CT - NO CONTRAST (risk of hyperviscosity). <p>Transfusion support:</p> <ul style="list-style-type: none"> • options include initial simple transfusion to Hb 100 g/L followed by red cell exchange. 	<p>Two forms – intermittent or prolonged.</p> <p>Specific management: Do not use ice.</p> <ul style="list-style-type: none"> • Simple measures e.g. moderate exercise, take a bath or shower • Empty bladder – may need catheter • Analgesia, oxygen, hydration with alkalinisation of the urine should be commenced as soon as possible. <p>Consult General Surgery and on-call haematology fellow if priapism has lasted more than 3-4 hours. Transfusion support.</p>	<p>An acute illness with a decrease in haemoglobin without a reticulocyte response (usually <1%). Usually associated with acute infection including parvovirus. Present with pallor +/- shock.</p> <p>Specific management:</p> <ul style="list-style-type: none"> • Intravenous fluids and oral intake to a total of maintenance • Transfuse red blood cells if patient is asymptomatic with anaemia or Hb <50 g/L (do not increase Hb by >30 g/L) • Commence IV antibiotics if febrile – Ceftriaxone. 	<p>Anaemia (↓Hb >20g/L) with thrombocytopenia and acute splenomegaly. May present acutely shocked.</p> <p>Specific management:</p> <ul style="list-style-type: none"> • Fluid resuscitation – sodium chloride 0.9% 10 – 20 mL/kg • Initial transfusion to aim for Hb of 50 - 60 g/L initially to ameliorate haemodynamic instability (do not increase > 30 g/L) <p>Auto-transfusion may occur if haemoglobin is increased excessively or too quickly. This increases risk of stroke due to hyperviscosity.</p> <p>IV antibiotics if febrile as per ChAMP guidelines.</p>

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