### Initial management of Sickle cell disease – ED pathway

#### Triage and assessment

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

Check for signs of complications, e.g.:
- 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:** Oxycodeone PO or morphine 0.05mg/kg IV, repeat as needed, may need continuous infusion.

**Fluids:**
- Push oral fluids
- May require IV fluid bolus 10 – 20 mg/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 PICU review and respiratory support if concern for acute chest crisis.

**Fever – sepsis**
- Consider cover for atypical organisms.
- Obtain appropriate cultures: blood, sputum, urine.
- Early referral to PICU if significant hypoxia or chest distress.

**Acute chest syndrome**
- Suspect if significant respiratory distress, hypoxia or chest pain.
- Oxygen to keep O2 saturation > 96% or for comfort.
- Intravenous fluids and oral analgesia.
- Chest X-ray – but this should not delay commencement of treatment.
- Early referral to PICU if patient is acutely shocked.

**Stroke**
- Early referral to PICU for respiratory support if concern for acute chest crisis.
- CT - NO CONTRAST (risk of haemorrhagic stroke).

**Priapism**
- May require blood transfusion – discuss with Haematology fellow.
- Consult General Surgery for haematology fellow if priapism has lasted more than 3-4 hours.
- Transfusion support:
  - Simple measures e.g. alkaclinisation of the urine.
  - Empty bladder – may need catheter.
  - Analgesia, oxygen, hydration with topical anaesthetic cream.

**Aplastic crisis**
- Usually associated with acute splenic sequestration.
- Anaemia (< Hb >20g/L) with thrombocytopenia and acute splenomegaly. May present acutely shocked.

**Acute splenic sequestration**
- This increases risk of stroke due to hyperviscosity.
- IV antibiotics if febrile as per Champs guidelines.

#### Contact Haematology Fellow on call

**Investigations:**
- FBC including reticulocyte count
- Blood group & cross match
- CRP, blood and urine cultures if febrile
- U&Es and LFTs if dehydrated
- Respiratory gases if breathing air
- MRIs if not available
- CXR if respiratory symptoms
- Other imaging required.

<table>
<thead>
<tr>
<th>Vaso-occlusive crisis (painful crisis)</th>
<th>Fever – sepsis</th>
<th>Acute chest syndrome</th>
<th>Stroke</th>
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<th>Acute splenic sequestration</th>
</tr>
</thead>
<tbody>
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<td>Precipitated by dehydration, hypoxia or infection. All episodes of pain should be treated initially as vaso-occlusive disease as per General Sickle Crisis Management. Chest pain may indicate an acute chest syndrome rather than as a vaso-occlusive episode if associated with respiratory symptoms.</td>
<td>Patients are functionally asplenic and at greater risk for invasive disease by encapsulated organisms. Specific management:</td>
<td>Commence IV Ceftriaxone Consider cover for atypical organisms (Azithromycin) if significant respiratory component Obtain appropriate cultures: o 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 O2 saturation &gt; 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 PICU for respiratory support if significant hypoxia or respiratory distress.</td>
<td>Can occur suddenly or as a complication of acute chest syndrome or aplastic crisis. Specific management: Neuroimaging required to determine if haemorrhagic or ischaemic stroke. MRI is modality of choice if not available CT - NO CONTRAST (risk of hyperviscosity). Transfusion support: options include initial simple transfusion to Hb 100g/L followed by red cell exchange.</td>
<td>2 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 alkaclinisation of the urine should be commenced as soon as possible. Consult General Surgery and on-call haematology fellow if priapism has lasted more than 3-4 hours. Transfusion support.</td>
<td>An acute illness with a decrease in haemoglobin without a reticulocyte response (usually &lt;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 anaemia or Hb &lt;50g/L (do not increase Hb by &gt;30g/L) Commence IV antibiotics if febrile – Ceftriaxone.</td>
<td>Anaemia (Hb &gt;20g/L) with thrombocytopenia and acute splenomegaly. May present acutely shocked. Specific management: Fluid resuscitation – 0.9% sodium chloride 10-20ml/kg Initial transfusion to aim for Hb of 50-60 g/L initially to ameliorate haemodynamic instability (do not increase &gt; 30 g/L) Auto-transfusion may occur if haemoglobin is increased excessively or too quickly. This increases risk of stroke due to hyperviscosity. IV antibiotics if febrile as per Champ guidelines.</td>
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