PAEDIATRIC ACUTE CARE GUIDELINE	
Sickle Cell Disease	
Scope (Staff):	All Emergency Department Clinicians
Scope (Area):	Emergency Department

This document should be read in conjunction with this DISCLAIMER
http://kidshealthwa.com/about/disclaimer/

Sickle Cell Disease

Sickle Cell Disease ED Pathway quick reference.

Background

Sickle cell disease (SCD) is a haemoglobinopathy where abnormal haemoglobin (HbS) "sickles" causing blood flow obstruction.

Sickle cell crises can be spontaneous or precipitated by:

- Dehydration
- Hypoxia
- Infection

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

Presenting problems

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

Evaluate for complications and begin urgent treatment after discussion with the

Oncology/Haematology Fellow on-call.

General Management

A child with SCD presenting to ED with fever or pain should be assessed **within 30 minutes** of arrival. Consult on call Haematology Fellow/Haematologist early.

Do **not** wait for EMLA for commencement of IV fluids or analgesia.

Analgesia

Start analgesics promptly:

- mild – paracetamol & ibuprofen
- moderate to severe – oxycodone oral or morphine 0.05mg/kg IV

Repeat as needed – may need opioid continuous infusion.

Fluid

- Push oral fluids
- May require IV fluid bolus 10-20ml/kg
- Consider maintenance IV if unable to tolerate oral
- **Avoid excess fluids** to reduce risk of chest crisis

Oxygen

- For hypoxia or respiratory distress
- Early PICU review and commence respiratory support as soon as possible if clinical concern of acute chest syndrome

Blood Transfusion

- May be required – discuss with on call haematology fellow/haematologist

Investigations

- FBC including reticulocyte count
- Blood group & cross match
- Blood and urine cultures if febrile CRP
- U&Es and LFTs if jaundice or dehydrated
- Consider chest Xray if febrile with respiratory symptoms

Consider other imaging as clinically appropriate

Specific Management

Vaso-Occlusive Crisis

Precipitated by dehydration, hypoxia or infection.
All episodes of pain should be treated initially as vaso-occlusive disease as per general management above.

NB Chest pain may indicate an acute chest syndrome rather than as a vaso-occlusive episode if associated with respiratory symptoms.

Fever

Patients are functionally asplenic and at greater risk for invasive disease particularly by encapsulated organisms (eg. Haemophilus, Meningococcus, Streptococcus).

Specific Management:

- Obtain appropriate cultures – blood, sputum, urine
- Commence IV antibiotics as per Champ guidelines
- Cover for atypical organisms (Azithromycin) if significant respiratory component
- Treat pain as per vaso-occlusive crisis
- Consider and treat for acute chest syndrome if cough or dyspnoea is present

Acute Chest Syndrome

Chest crisis is a life threatening condition and patients can deteriorate rapidly.

Suspect if respiratory distress, hypoxia or chest pain

- Oxygen to keep O2 saturation > 96% or for comfort
- Analgesia
- Commence IV antibiotics – Ceftriaxone and Azithromycin
- Chest Xray – but this should not delay commencement of treatment
- Early referral to PICU for respiratory support if significant hypoxia or respiratory distress

Stroke

Incidence is up to 1 in 10 patients with HbSS disease.
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 preferred
- CT without contrast (risk of hyperviscosity) if MRI not available

Transfusion support:

- Discuss with on call Haematology Fellow/Haematologist
- Transfusion to Hb 100g/L +/- red cell exchange

Priapism

May be intermittent or prolonged (>4 hours increases risk of impotence)

Specific Management:

- Do **not** use ICE
- Analgesia, oxygen, IV hydration
- Empty of bladder – may need catheterisation
- Simple measures eg. moderate exercise, take a bath or shower
- Alkalanise urine
- Consult on-call Haematology Fellow/Haematologist and General Surgery if priapism has lasted more than 3-4 hours
- Transfusion support may be required – discuss with Haematology

Aplastic Crisis

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

Specific management:

- Intravenous fluids and oral intake to a total of maintenance
- Transfuse red blood cells if patient is symptomatic with anaemia or Hb <50g/L (do not increase Hb by > 30g/L)

- Commence IV antibiotics if febrile – Ceftriaxone

Acute Splenic Sequestration

Anaemia (drop in Hb >20g/L) with mild to moderate thrombocytopaenia and acute splenomegaly. May have co-existent aplastic anaemia if reticulocyte count is low. May present in shock.

Specific management:

- Fluid resuscitation – 0.9% sodium chloride 10-20ml/kg for hypovolaemia while waiting for blood
- Initial transfusion to aim for Hb of 50-60 g/L initially to ameliorate shock/haemodynamic instability (do not increase > 30 g/L)

Autotransfusion will 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

Tags

aplastic, chest crisis, priapism, sickle, vaso-occlusive

This document can be made available in alternative formats on request for a person with a disability.

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