

Clinical Guidance

Paediatric Critical Care: Emergency management Sickle cell disease (SCD)

Summary

This guideline outlines the emergency management of patients with sickle cell disease presenting with acute complications and indications for tertiary HDU/PICU admission. There are other guidelines for sickle cell available covering specific aspects of Sickle cell- see links below for four Evelina documents.

Document Detail	
Document type	Clinical Guideline
Document name	Paediatric Critical Care: Emergency management Sickle cell disease (SCD)
Document location	GTi Clinical Guidance Database
Version	V 1.0
Effective from	30 November 2023
Review date	30 November 2026
Owner	Head of Service, PICU
Author(s)	Shelley Riphagen (PICU Consultant)
Approved by, date	Evelina London Clinical Guideline Committee
Related documents	Pan-London SCD Escalation Guideline ; Evelina Internal sickle guidance: Paediatric: Red Blood Cell Exchange Transfusion (RBCX) , Priapism , Fever, pain ,
Keywords	Sickle, Sickle Cell Disease, PICU, Paediatric, Emergency, STRS, Acute Chest Syndrome, Crisis, Genetic disorder, Transfusion, Exchange.
Relevant external law, regulation, standards	

Change History		
Date	Change details, since approval	Approved by

Paediatric Critical Care

Emergency management Sickle cell disease (SCD)

Sickle cell disease (SCD) Autosomal recessive genetic disorder producing HbSS.1:2000 births in England. Neonatal screening. Symptoms start after disappearance of HbF at 4-6 months

Emergencies

Refer to Specialist Haemoglobinopathy Team, agree treatment pathway and optimal location. Involve STRS if transfer required.

- **Acute chest syndrome (ACS):** Clinically and radiologically indistinguishable from pneumonia so both should be treated. Associated with higher mortality. Dyspnea, fever, chest/ upper abdominal pain, cough, tachycardia, tachypnea, wheeze, hypoxia (sats < 95% in air). Children may deteriorate rapidly. Escalate care appropriately.
- **Acute girdle syndrome:** Circumferential severe abdominal pain. Acute vaso-occlusion (splenic/ hepatic sequestration), infective or unrelated cause.
- **Acute neurologic episodes:** Refer any acute neurology in SCD for specialist advice. Transient ischaemic attack, seizure, overt stroke (250x more common in SCD) or acute visual impairment (plus urgent ophthalmology review)
- **Severe sepsis:** Increased risk of osteomyelitis, cholecystitis
- **Priapism:** Urology referral. Intra-cavernosal aspiration/ washout/ Water's shunt
- **Hyper-haemolysis syndrome.** Refer to tertiary sickle cell disease. Discuss case with local haematology/ blood transfusion service urgently
- **Splenic sequestration:** new splenomegaly with anaemia (also caused by aplasia)
- **Severe liver dysfunction:** Biliary sepsis, hepatic sequestration, acute hepatitis. Urgent hepatology & haematology input. Urgent ERCP/ MRCP

Medical management

- Maintain **oxygen saturations >95%**. Incentive spirometry.
- Low threshold for **respiratory support** with humidified high flow nasal cannula oxygen or CPAP (PEEP 5-7). Should only be set up with agreed escalation plan for transfer to HDU/ PICU
- **Bronchodilators** may be useful in SCD with associated wheeze
- Initiate **adequate pain relief** with paracetamol, NSAIDs and opiates if necessary. Rapid acting intranasal diamorphine or fentanyl while IV access secured. Pain can be severe and control difficult. **Reassess pain every 30 mins** and **escalate management** until relief achieved.
- Obtain **two good peripheral IV access**. Escalate to experienced operators early, as may be challenging
- Identify and resuscitate **shock**. Consider **blood products** early as **volume resuscitation**. Be aware allo-antibodies possible.
- After **blood culture**, use **appropriate antibiotics**- IV cephalosporin and macrolide as per local policy. Stop penicillin prophylaxis.
- If Hb <70g/L or below own baseline, **transfuse to 90-100g/L**. Allo-antibodies common. Discuss with haematology prior to transfusion. Monitor closely.
- **Strict fluid balance**. Fluid restrict to 75% maintenance for those on non-invasive respiratory support. Restrict to 50% when invasively ventilated. Avoid diuretics in acute presentation, except on specialist advice.
- **Suspected stroke**. ABCD as above. Urgent Brain CT with contrast within 1 hour. Top-up transfusion if Hb<80g/L. For exchange transfusion if stroke. If intracranial haemorrhage refer to neurosurgical centre.

Indications for tertiary HDU/ PICU admission

Early referral to PICU retrieval team with urgent transfer

- Escalating oxygen and respiratory support requirements
- Already deployed non-invasive ventilation
- Acute stroke or other organ failure
- High HbS % (>30%) and acutely unwell. Will possibly need exchange transfusion

For intubation and ventilation if

- Worsening hypoxia or dyspnea
- Worsening respiratory acidosis despite NIV
- Depressed level of consciousness

Resources: [Pan-London SCD Escalation Guideline](#)

Royal London 0207 377 7000
Evelina London Children's 0207 1887188
Kings College 0203 299 4000
St Mary's 0203 312 6666
Great Ormond Street 0207 405 9200
Paediatric emergency referral and retrieval
CATS 0207 430 5850
STRS 0207 188 5000

Emergencies arise due to

- Sepsis
- Vaso-occlusive episodes due to sickling
- Aplastic crises induced by parvo virus
- Sequestration episodes
- Acute haemolytic crises
- Need for emergency surgery

Precipitants include:

Hypoxia, infection, dehydration, acidosis
Stress and anxiety, pain, extreme hot or cold environment, extreme exercise and episodes of "spontaneous" vaso-occlusive crisis (VOC)

Special investigations

- Establish child's usual baseline Hb and oxygen saturations
- Perform FBC with reticulocyte count
- HbS (sickle) percentage.
- Blood gas, lactate and glucose
- Electrolytes, renal and liver function
- Blood culture
- Cross match (Request sickle negative, Rh and Kell matched as minimum)
- Send NPA with viral & atypical serology
- Perform CXR & abdominal ultrasound
- Ferritin level (chronic transfusion therapy)
- Urgent CT head if neurologic symptoms

Special considerations

- **Exchange transfusion may be necessary.** Must be undertaken in fully monitored HDU conditions
- **Plastic bronchitis** presents as wheezing, poorly or incompletely responsive to bronchodilators. Refer for? bronchoscopy.
- **Delayed haemolytic transfusion/ hyper-haemolysis** may occur due to allo-immunisation; presents with shock and hyperkalaemia.

Exchange transfusion in SCD

Consider in management of acute emergencies

- **Aim:** HbS<30%, Hb >100g/L, Hct < 35%
- **Pre-considerations:** Hb<70g/L – give top up exchange prior to partial exchange.
- Transfused blood <10 days old & fully matched
- Partial exchange transfusion **volume:** 20mL/kg
- **Location:** Fully monitored in HDU/ PICU with nurse and doctor/ advanced nurse practitioner
- **Technique:** Use one cannula to withdraw blood & one to infuse equal small aliquots equal volume blood+ balanced crystalloid.
Record all input and removal. **Check** electrolytes, glucose and gas half way through procedure.

References

BMJ best practice(2018) Sickle cell anaemia. Lancet (2017) Ware NHLBI (2014) Evidence based management of SCD SCD is childhood: Standards & recommendations 2019 Pan-London SCD Guideline 2022