

GSTFT Clinical Practice Guideline

**Guidelines for the Initial Management of Children with
Sickle Cell Disease (SCD) in A&E**

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Guidelines for the Initial Management of Children with Sickle Cell Disease (SCD) in A&E

SCD is a chronic condition affecting many different organs of the body and is also characterised by acute episodes; the commonest being the painful episode, some acute complications are life-threatening and not always associated with pain. They may be insidious in their presentation eg:

Pneumococcal sepsis:	Symptoms may be few with sudden deterioration
Acute splenic sequestration:	Hb may fall rapidly - always measure the spleen.
Cerebrovascular accident:	Minor symptoms eg paraesthesiae may herald CVA
Acute chest syndrome:	Often associated with pain in trunk/girdle area: fall in O ₂ sats, change in bedside observations and chest signs may precede X-ray changes.

General Principles when Assessing Children with SCD:

It is important to be aware of previous history and results so check EPR for steady state investigations and correspondence. A few children have individual care plans which they should bring with them (copies should be available in A&E.)

• Pain (see also pain guideline)

Children can become very anxious when in pain, so please comfort and try to reassure. Assess for possible infective triggers but clinical signs may be minimal in a painful episode. If diverted, the child's pain may seem to lessen only to return when a parent is present; this child is unlikely to be 'putting on' his symptoms but it makes it difficult to assess.

Osteomyelitis is extremely rare. X-rays and isotope scans in the early stages are not helpful. If there is a strong clinical suspicion of osteomyelitis (localised pain, swelling, high fever, systemically ill child), request ultrasound and consult the Paediatric Sickle Cell team, and Paediatric Orthopaedic team to be contacted as ultrasound is requested.

• Fever (see also fever guideline)

Children with SCD are hyposplenic and at risk of overwhelming sepsis, particularly from pneumococcus, meningococcus, haemophilus and salmonella. There may be very few clinical signs, and a child with fever needs a septic screen (blood + urine culture, atypical serology, viral serology including parvovirus) and close monitoring. However sepsis is very rare as children are usually on prophylactic penicillin and/or have been immunised. It is very common to be febrile with a painful episode. Only start empirical antibiotics (cefuroxime iv and erythromycin po) if child unwell or temperature $\geq 38.0^{\circ}\text{C}$ after baseline investigations have been taken. In other children remember to continue their prophylactic penicillin V. Remember to check on blood results and compare with steady state values. Consider malaria depending on travel history.

• Dehydration

Children with SCD are hyposthenuric and if vomiting or not taking adequate oral fluid give iv fluids (see below for amounts). A creatinine result in the high normal range in a child with SCD is significant.

• Anaemia (see also low Hb guideline)

Mild anaemia may be difficult to detect clinically but it should be possible to detect severe anaemia. Check the haemoglobin of all children with SCD at triage. Always palpate for the spleen, measure and compare with previous values. See below for investigations and transfusion policy. Reassess at least daily and monitor the Hb and reticulocyte count.

History Taking & Documentation on Admission

Some children are admitted frequently - it may not be appropriate to cover all the points below.

- History: present episode (not all SCD complications are associated with pain and not all medical or surgical problems will be caused by SCD).
- Pain: record site(s), assess severity using paediatric early warning signs (PEWS) and pain score. Record analgesic dosages and times.
- Previous medical history: SCD & other problems, chest crises/admissions/time off school. Ask if the spleen is usually enlarged.

- Sickle cell phenotype: most parents know this, check on EPR, with red cell lab, or in the notes. Please use the following convention: HbSS (sickle cell anaemia), HbSC (Haemoglobin SC disease), HbSBeta Thal or HbSBeta Thal (Haemoglobin sickle beta thalassaemia).
- Steady state Hb: Ask parent or check EPR/notes.
- Penicillin V: compliance.
- Prevenar/Pneumovax immunisation history (or check EPR under summary) : If new to King's take a full history.
- Known allergies.
- History of transfusions and any reactions.
- Travel History/malarial prophylaxis
- Usual hospital for follow-up, date of last OPA and when next to be reviewed.
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Initial Investigations –

Send all blood tests at first venepuncture to avoid repeated sampling

- FBC & retics. If Hb <5g/dl or ~2g/dl below steady state, consider transfusion. The reticulocyte count is markedly raised in splenic sequestration, and zero/very low in aplastic crisis. Request parvovirus serology if concerned and isolate because of risk to other patients and pregnant women.
 - DAX + ALT + LDH, (and amylase if abdominal pain).
 - Group & Screen
 - Septic screen: MSU, blood cultures, virology sample and consider LP, malaria screen.
 - CXR, abdominal U/S as indicated.
 - Pulse oximetry if respiratory sign/symptoms or on opiates. Compare with steady state.
 - Other investigations eg MRI scans in stroke as indicated, stool cultures if salmonella osteomyelitis suspected.
- NB** There is no value in measuring the sickle percentage (HbS%) unless transfused < 3/12 before, or following exchange transfusion.

Initial Treatment:

Some children, particularly those who attend frequently will have a care plan in A/E

- **Analgesia:** check what has already been given at home and see separate protocol for pain relief. Please remember to prescribe regular laxatives.
- **Antibiotics:** consider if febrile >38.0°C (see above). Start iv cefuroxime and oral erythromycin following cultures. Continue prophylactic penicillin V if no other antibiotic prescribed (<1yr 62.5mg po bd, 1-5 years 125mg po bd, >5 years 250 mg po bd).
- **Fluid:** Children should be appropriately hydrated .Requirements may be greater than normal as most SCD children are hyposthenuric. However it is important not to over hydrate eg if chest signs. Encourage oral fluids but if not tolerated give iv 0.18% NaCl/4% Dextrose and monitor fluid balance & electrolytes.

Maintenance iv fluid requirements are as below but some children may require more.:

Age (years)	Normal maintenance (ml/kg/24hrs)
1-3	100
4-6	90
7-14	70
15-18	60

- **Blood Transfusion** - see Trust Policy and Guidelines on Cliniweb for full details.

If you think a child needs blood transfusion always discuss with the paediatric haematology team. Phenotypically-matched, sickle-negative red cells should be used. Request CMV negative blood if CMV status is unknown or negative.

Simple (top-up) transfusion is indicated in aplastic crisis, splenic sequestration, chest symptoms with a fall in Hb or maintenance if the child is on regular transfusion. Do not exceed a PCV of 0.35, Hb 8 -10g/dl.

Exchange transfusion is indicated for acute CVA, chest syndrome, and life-threatening infection; the target HbS <30% and PCV <0.35. Two exchanges may be needed to achieve this.

Pre-Operative Transfusion (and see Guideline for Paediatric SCD patients undergoing anaesthesia)

Minor procedures: Ts & As, circumcision, hernia, appendectomy, possibly cholecystectomy – Group and Antibody Screen

Major surgery: joint replacement, organ transplant, major abdominal surgery: Top-up transfusion to Hb 10 g/dl and document HbS% or partial exchange if significant previous medical history e.g. sickle chest.
Complex major thoraco-abdominal or neurosurgical procedures: exchange transfusion to reduce HbS% to <30%, PCV <0.35.

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