

The Royal College of Emergency Medicine

Best Practice Guideline

**Management of Acute
Presentations of Sickle
Cell Disease in the
Emergency Department**

Summary of recommendations

1. All Emergency Departments should be aware of sickle cell disease, be able to recognise cases and have knowledge of the potential complications.
2. All patients presenting acutely with sickle cell disease should be triaged urgently, aiming to assess within 30 minutes.
3. All Emergency Departments should have a locally agreed care pathway for the management of sickle cell disease.
4. Recognition and alleviation of pain should be a priority. All patients should be assessed and offered analgesia if required within 15 minutes of arrival, and reassessed within 15 minutes if in severe, or 30 minutes if in mild-moderate pain.
5. Ensure that all acutely unwell sickle cell patients are cared for in an appropriate and warm environment, or have warming measures provided (such as a Bair Hugger), with provision of drinking water.
6. Initial assessment and decision regarding the identification of a sickle cell disease complication or severe pain crisis should occur within one hour of presentation.
7. Decision-making for uncomplicated cases regarding discharge or appropriate disposition such as a haematology day care unit or agreed Same Day Emergency Care pathway (where available) should be made within two hours.
8. Identification of a significant sickle cell disease complication or severe pain crisis warrants prompt discussion with haematology services and prompt referral and discussion with an admission team such as critical care, if required. Such patients should be referred and seen by the relevant speciality within 30 minutes of recognition of the potential complication.
9. If specialist haematology services are required and cannot be provided on-site, the patient will require urgent transfer of care to a centre with appropriate facilities and expertise. Transfer agreements with the local ambulance service should be in place with an agreed pathway and effective communication process.
10. All decisions concerning treatment and transfer should, wherever possible, be made in conjunction with the patient and/or their carer/family.

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Scope of the Guideline

This guideline has been developed to provide recommendations on the initial care of patients presenting to the emergency department (ED) with acute sickle cell disease (SCD) presentations and its potential complications. The aim is to increase Emergency Department (ED) awareness of SCD, optimise and standardise the management of such patients, and ensure equity of access to acute care and specialist services.

The process of creating this guideline has included contemporaneous literature reviews for high-level evidence in the medical literature on all aspects of sickle cell disease, as well as searches for consensus agreement publications where they exist. The guideline group have formed consensus on areas lacking clear answers.

Reasons for Development

An All Party Parliamentary Group (APPG) report entitled 'No One's Listening' was published in November 2021 highlighting avoidable deaths and failure in care for SCD patients in secondary care. The report acknowledged that the care of those with SCD has been low priority and received insufficient funding for decades. Racial bias and stigmatisation of predominantly young black individuals living with SCD has been recognised. Patients presenting in extreme pain are often accused directly or indirectly of drug seeking and report high levels of anxiety and a lack of empathy and respect when accessing emergency care. A Health Service Safety Investigations Body (HSSIB) report from June 2023 also included recommendations regarding training of staff in the awareness of SCD, and recognition and management of potential sickle cell crisis.

Introduction

Sickle cell disease (SCD) is a group of lifelong inherited haemoglobin abnormalities. More than 15,000 people live with SCD in the UK, with increasing prevalence due to immigration and new births. Approximately one in 79 babies carry the sickle cell trait, and around 300 babies are born with sickle cell disease each year. Most affected individuals are of African or African Caribbean heritage, although it can affect anyone.

SCD has a significant impact on morbidity and mortality in affected individuals. Emergency physicians, acute medicine and haematology services should work collaboratively to develop local care pathways for SCD, and to encourage and support training and education regarding the condition for clinical teams.

Pathophysiology and Potential Complications of SCD

Acute vaso-occlusive episodes (VOE), also known as vaso-occlusive crises, or painful sickle crises, are caused by blockage of capillaries by sickled cells, with subsequent tissue infarction. In SCD, the usually flexible biconcave red blood cells can become rigid and 'sticky' as a result of polymerisation of sickle haemoglobin. Precipitants for red cell sickling include dehydration, hypoxia, and infection. Repeated episodes of ischaemia result in end organ damage. Many individuals with SCD have some form of significant organ impairment, which includes sickle nephropathy, sickle hepatopathy, sickle retinopathy, stroke, reduced lung function, pulmonary hypertension, hyposplenism, avascular necrosis particularly of the hip and shoulder, priapism and erectile dysfunction, chronic leg ulceration, and osteopenia.

VOE can occur unpredictably, sometimes without clear precipitating factors, and with marked variation in frequency, duration, and severity. Most pain episodes are managed at home and patients usually present for hospital management only if their pain is uncontrolled or they have no access to analgesia.

The primary goal in the management of a VOE is to achieve effective prompt and safe pain control. Unacceptable delays to analgesia, insufficient or excessive doses, inappropriate analgesia, and stigmatising the patient as drug seeking are commonly reported.

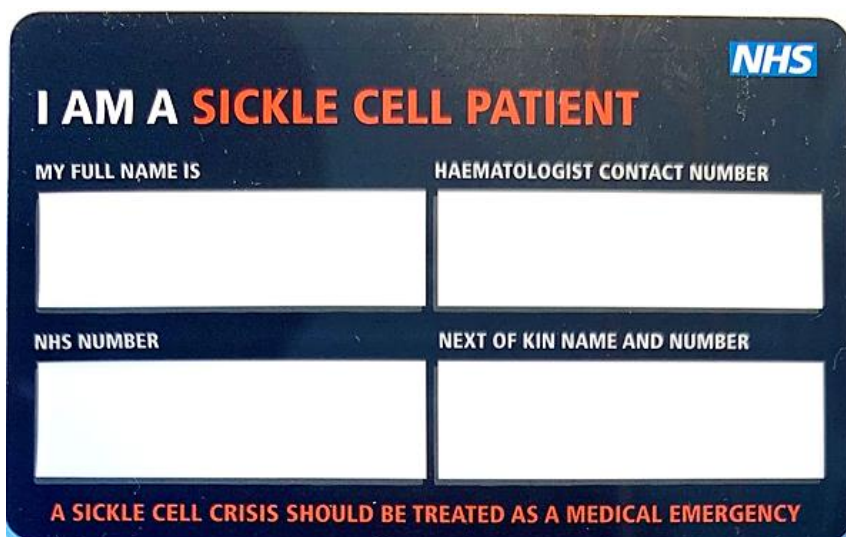
Local acute pain management protocols for those with SCD, and, especially for adults, **individualised care plans**, should be readily available and include medication choices and advisory dosages, taking into account patients' needs and preferences.

Recognition and Triage

It is important to remember that SCD can affect anyone. However, it mostly affects individuals from black African and Caribbean backgrounds. In addition, people of Central and South American, Middle Eastern, Mediterranean and South Asian heritage may also be affected.

An acute painful sickle cell episode is an acute medical emergency. All cases should be triaged as high priority and assessed in a 'main' ED area, with access to appropriate monitoring and trained staff.

Patients may carry an emergency card which looks like this:



The image shows a dark blue NHS emergency card. At the top right is the NHS logo. The main heading is 'I AM A SICKLE CELL PATIENT' in white and orange text. Below this are four white rectangular boxes for information: 'MY FULL NAME IS', 'HAEMATOLOGIST CONTACT NUMBER', 'NHS NUMBER', and 'NEXT OF KIN NAME AND NUMBER'. At the bottom, a red banner contains the text 'A SICKLE CELL CRISIS SHOULD BE TREATED AS A MEDICAL EMERGENCY'.

Initial Management of Acute Pain

NICE has published guidelines on the management of acute sickle pain and below is a summary. Refer also to the summary in Appendix 1.

The patient (and/or carer) is an expert in their condition. Listen to their views and take into account:

- treatment received during previous episodes
- any concerns they have about the current episode
- any psychological and/or social support required.

Assess pain with an age-appropriate pain severity scoring tool at presentation.

Offer analgesia within 15 minutes of presentation. Re-evaluation should occur within 15 minutes for severe pain and within 30 min for minor-moderate pain.

In patients who are naïve to regular/ strong opiates, effective analgesia can often be achieved with a combination of non-steroidal anti-inflammatory drugs (NSAIDs), paracetamol and then consider adding codeine, dihydrocodeine, or oxycodone if required.

If this is not successful, or the pain at presentation is severe, offer a bolus of opiate analgesia as per their individual care plan or, if there is no individual care plan available consider morphine at the following dose:

- **For adults (≥16 years): morphine 0.05 - 0.1 mg/kg S/C**, repeated every 30 minutes until the pain improves and provided no adverse effects (excessive sedation, respiratory depression), after which 0.05 - 0.1 mg/kg 2-4 hourly S/C can be used as maintenance. The IV route is often reserved for when transfusions or parenteral antibiotics are required, as access can often be difficult to obtain.
- **For children: morphine 0.2 - 0.3 mg/kg PO (max 10mg) or 0.1 mg/kg IV (max 5mg) as a bolus**, repeated every 30 minutes until the pain improves and provided no adverse effects (excessive sedation, respiratory depression), after which consider 0.2 - 0.3 mg/kg PO (max 10mg) 2-4 hourly or an IV/SC patient/nurse controlled analgesia regime (follow local policy on administration/ monitoring).

Clinical Assessment

Clinical assessment of all patients presenting to the ED with an acute painful sickle cell episode should include initial and, ideally, continuous (minimum half-hourly) monitoring of:

- Blood pressure
- Oxygen saturation (SpO₂) - *offer O₂ therapy if SpO₂ is 95% or below*
- Pulse rate
- Respiratory rate
- Temperature
- NEWS/ PEWS

Additionally, also measure the child's weight on arrival.

ESCALATE PROMPTLY if:

- **(P/N)EWS ≥4, or recorded as 3 in any category**
- **SpO₂ < 94% on air, or there is an increasing oxygen requirement.**

Assessment must determine whether pain is being caused by an acute VOE or an alternative diagnosis, particularly if the patient reports atypical pain.

When offering analgesia:

- Appreciate whether the patient is opiate naïve

- Record and take into account analgesia already taken for the current episode before presentation
- Ensure drug, dose and administration route are suitable for pain severity and patient's age
- Refer to the patient's ***individual care plan***, if available.

Offer a bolus dose of strong opioid by a suitable administration route, to:

- All patients presenting with severe pain
- All patients presenting with moderate pain who have already had analgesia before presentation.

Boluses of opiate analgesia are usually of morphine or oxycodone subcutaneous or oral equivalent with the dose varying depending on the individual's prior opiate exposure and tolerance (e.g. morphine 2.5-15mg s/c or oxycodone 2.5-10 s/c 2-4 hourly). In the paediatric setting, intranasal fentanyl can be an alternative to IV/SC if access or severe needle phobia is an obstacle.

If severe pain persists despite supportive measures and three boluses of strong opiate analgesia, then it may be appropriate to consider the use of subcutaneous Patient Controlled Analgesia (PCA) with either morphine or oxycodone (e.g. Oxycodone 2mg bolus, or paediatric adjusted dosing, maximum every 10 minutes). Local guidance should be followed and training for staff who may care for SCD patients with a PCA is essential.

Due to risks regarding opiate toxicity, PCA is usually restricted to wards with expertise in their use, such as a haematology ward or HDU. To reduce the risk of opiate toxicity, most PCA protocols are initiated without a background infusion. However, slow release oral opiates can be prescribed in addition if required, with the dose varying depending on the patient's prior opiate exposure and tolerance.

Where the patient has known side effects or prior nausea/ vomiting, an antiemetic may be offered prior to administration of an opiate bolus. First line options include ondansetron, prochlorperazine, or metoclopramide. Metoclopramide should not be first line in young and particularly female patients due to possible dystonic side effects. IV/SC cyclizine boluses should be avoided, but when parenteral antiemetics are required for patients unable to tolerate oral doses, cyclizine may be given by intravenous infusion over one hour.

Supportive care

Offer laxatives, antiemetics, antipruritics, paracetamol and NSAIDs in addition to an opioid, unless contraindicated. Avoid NSAIDs during pregnancy, unless the potential benefits outweigh the risks. NSAIDs should be avoided altogether in the third trimester.

Offer supportive care as outlined below:

- Oxygenation – maintain $\geq 95\%$ with supplemental oxygen
- Anti-emetic (e.g. metoclopramide)
- Regular oral analgesia (e.g. paracetamol; ibuprofen, codeine or slow release opiate)
- Anti-pruritic (e.g. chlorphenamine or hydralazine)
- Hydration: aim for euvolaemia orally; IV if required
- Observation, pain & NEWS score at 30 minutes then hourly for 1-6 hrs, then 4 hourly
- Antibiotics – if fever or signs of infection; noting higher-risk cases e.g. hyposplenism (see below)

- Thromboprophylaxis in adults, unless contraindicated
- Laxatives when on opiate analgesia or concerns regarding constipation

Encourage individual coping mechanisms (e.g. distraction and relaxation techniques) for dealing with acute pain.

Pethidine is not recommended for analgesia in an acute painful sickle cell episode (NICE).

Naloxone should be readily available for reversal of potential opiate toxicity.

Individual care plans allow a pre-agreed care plan to be followed from the moment the Ambulance Service attends the individual in their home and has the potential to avoid delays to effective analgesia and facilitate urgent transport to the patient's nominated centre.

Reassessment

Assess the effectiveness of pain relief:

- Every 15-30 minutes until an acceptable level of improvement in pain has been achieved and at least every 4 hours thereafter based upon the patients reporting, or using an age-appropriate pain scoring tool or visual analogue score (VAS).
- Avoid unconscious bias or stigmatisation by asking questions, such as:
 - How well did that last painkiller work?
 - Do you feel that you need more pain relief?

If pain is indicated as severe (e.g. score >7) on reassessment, a second bolus dose of a strong opioid (or a first bolus dose if they have not yet received a strong opioid) should be offered. There is no requirement to wait two hours before repeating an opiate dose, provided there is no clinical evidence of opiate toxicity.

Ongoing management with a PCA may be indicated if repeated bolus doses of a strong opioid are required within the initial 2-hour period. Specialist advice will likely be needed in this case and warrant inpatient admission to provide this. It is unlikely that the PCA will be commenced in the ED.

Monitor patients taking strong opioids for adverse events, and perform a clinical assessment (including sedation score):

- every hour for the first 6 hours
- at least every 4 hours thereafter

If the patient does not respond to standard treatment for an acute painful sickle cell episode, reassess for a potential alternative or additional diagnosis. Non-pharmacological, approaches such as massage, heat pads and psychological/relaxation techniques can be beneficial.

As the vascular occlusive episode starts to resolve, step down pharmacological treatment, in consultation with the patient.

Uncomplicated SCD presentations

Uncomplicated acute pain episodes can potentially be diverted to a suitable Same Day Emergency Care (SDEC) setting or speciality day unit, if available. Such units are often restricted to daytime hours and may only take ambulant patients without signs of acute chest syndrome, priapism or non-sickle pathology (e.g. gastrointestinal haemorrhage).

In adults, if pain settles after two opiate doses and the patient is clinically stable, utilise a shared decision making approach with the patient and/or carer to consider discharge after 4 hours with an appropriate follow-up plan, analgesia management with opiate stewardship, safety-netting advice and provide emergency contact details.

Consider a lower admission threshold for children or pregnant patients requiring ongoing strong opiate analgesia.

Complicated SCD presentations

Situations suggestive of a more complex SCD presentation requiring ongoing care and admission include:

- Fever >38°C
- O₂ on room air <95%
- Chest pain
- Unresolved priapism
- Ongoing severe pain
- New neurological symptoms

Investigations

- Full blood count (FBC) & reticulocyte count (an indicator of rate of red cell production),
- Renal & hepatic chemistry
- C-reactive protein (CRP)
- Group and screen (G&S)

In addition, consider the following based upon the presenting clinical features:

- Blood cultures (BC)
- Mid-stream urine culture microscopy and sensitivity (MSU),
- Sputum for culture, microscopy and sensitivity (sputum MC&S),
- Respiratory virus PCR including COVID, influenza A and B; others e.g. RSV
- Chest X ray (CXR)
- Arterial or venous blood gases (ABG/ VBG) Note: ABG should ideally be performed on room air, provided the patient can tolerate a period off oxygen support to achieve this

Other possible complications that can co-exist or develop following an acute painful episode include:

- Acute stroke
- Aplastic crisis
- Infections
- Priapism
- Osteomyelitis

- Splenic sequestration

Prompt discussion with specialist teams is required, referrals should be seen within 30 minutes.

Effective Communication

Escalation

Discuss promptly with Haematologist and admitting team and consider specific SCD complications (*further details below*). Patients with complicated VOE can deteriorate quickly. Consider liaising with critical care services early.

Those aged 16 to 18 years are likely to require admission to an adult ward depending on local agreements, bed availability and status regarding an individual's transition from children's to adults services. A paediatric ward should be considered, if available, for teenagers who have not transitioned to adult services.

If the ED site is separate to the haematology or general medical admission site, urgent ambulance transfer to the appropriate ward is required.

Women who are pregnant with a gestation of more than 16-23 weeks are likely to be more appropriately managed on an obstetrics ward, but local agreements on gestations deemed appropriate for obstetric admission vary.

Engaging with local haematology services

Particular situations to liaison promptly with local haematology services include:

- Acute Stroke
- Acute Chest Syndrome
- Acute anaemia
- Severe, persistent priapism
- Pregnancy

Specific Complications

Acute Chest Syndrome (ACS)

ACS can develop rapidly in patients with a VOE and can result in fatal outcome. Escalate urgently to a senior clinician if any of the following are present:

- Abnormal respiratory signs and/or symptoms
- Chest pain
- Fever $>38^{\circ}\text{C}$
- Signs and symptoms of hypoxia e.g. $\text{SpO}_2 <95\%$ or an escalating O_2 requirement.

ACS is defined as the presence of new pulmonary infiltrates on chest radiograph in combination with respiratory symptoms and/or fever in a patient with sickle cell disease. Clinical features include:

- Chest pain (sometimes pleuritic)
- Cough (may be productive)
- Shortness of breath/ tachypnoea
- Hypoxia

- Fever
- Wheeze
- Tachycardia

Chest signs include crepitations, bronchial breath sounds, and reduced air entry on auscultation or rhonchi with dullness to percussion. In the early stages, chest examination may be normal.

The following investigations should be carried out in ED:

- Urgent chest radiography - focal consolidation, bilateral multi-lobe opacities or lower lobe involvement occurs most commonly. Radiology changes may lag behind clinical findings so ACS should still be considered even in the absence of acute radiological changes.
- Arterial blood gases performed, ideally on room air.
- Full blood count, reticulocyte count, serum biochemistry, LDH, clotting screen, HbS%, CRP.
- Group and antibody screen, cross-match if necessary.
- Blood cultures.

The following investigations are important, but can usually wait until inpatient admission:

- Nose and throat swabs for respiratory viruses
- Sputum MC&S and viral PCR/culture if able
- Atypical serology (repeat after 3 weeks)
- Urine for Pneumococcal and Legionella antigens

The patient must be discussed immediately with a Consultant or at least ST3 level in Haematology and early liaison with High Dependency /Intensive Care Unit as required, as some patients will require ventilatory support.

Vital signs, the NEWS2, and pain score should be assessed at least hourly. Continuous SpO₂ monitoring is desirable.

Haematology advice may include early blood transfusion. The degree of hypoxia and respiratory compromise will govern the need for, and mode of, any blood transfusion. The approach of administering a 'top up' transfusion may be of benefit early in the course of acute chest syndrome. Prompt transfusion often averts the need for red cell exchange by improving tissue oxygenation. Timing of transfusion can be more important than a target haemoglobin S%. However, due to increased viscosity of sickle blood, the target for post transfusion haemoglobin should be no greater than 100g/L, unless the HbS% is less than 30%. Individuals with haemoglobinopathies require extended phenotype red cells for transfusion and may have red cell alloantibodies, making selection of suitable red cells for transfusion potentially challenging. A valid G&S sample, prompt request for cross matching, and liaison with a haematologist is required. Rarely, patients may require centrally stored frozen units or pre-treatment to avoid severe transfusion reactions.

Intravenous fluids should be given with care to avoid fluid overload, and a fluid balance chart maintained.

Adequate analgesia is recommended to prevent splinting of the diaphragm, hypoventilation, atelectasis, and hypoxia which can result in increased sickling. Due consideration must be given to the optimal safe use of opiates.

Non-steroidal anti-inflammatory drugs and low molecular weight heparin such as enoxaparin 20-40mg SC daily (according to weight and renal function) are recommended in the absence of contraindications.

Nebulised bronchodilators may be beneficial in the presence of reactive airway signs or if there is a past medical history of asthma.

Differential diagnoses include community acquired pneumonia (CAP) or pulmonary embolism (PE) should be considered, particularly if there is an ongoing requirement for supplemental oxygen. For antibiotic therapy, consider covering atypical organisms.

Acute neurological complications

New neurological symptoms need urgent investigation. Stroke is one of the most critical complications, resulting in significant morbidity and mortality. It can affect young children as well as adults.

Ischaemic strokes and intracranial haemorrhage require urgent discussion with stroke/ neurological services, depending on local arrangements, and Haematology teams. Urgent neuroimaging is recommended, according to local protocols e.g. CT brain +/- CT angiography.

The management of neurological complications is complex and should be provided jointly by haematology and stroke teams working in conjunction with each other. Initial treatment that may be considered includes urgent exchange transfusion. Reperfusion therapy with intravenous thrombolysis may be considered in acute ischaemic stroke in adult patients with homozygous SCD, despite a risk of haemorrhage, often associated with specific pre-existing vasculopathy. It is not thought to be of benefit in children with SCD and acute ischaemic stroke.

Infection and Sepsis

Pneumococcal & other infections are more common in those with SCD because of hyposplenism – the spleen is often autoinfarcted by early childhood. A septic screen should be completed and appropriate antibiotics started according to likely focus and local microbiology guidance.

Patients with CAP or ACS should start IV antibiotics, with additional cover for atypical organisms, according to local guidelines. For example, in adults, IV co-amoxiclav 1.2g tds with oral clarithromycin 500 mg bd, unless contraindicated. Consult the children's BNF for paediatric adjusted doses of these antibiotics. Ceftriaxone can be associated with haemolysis and is therefore not first line.

If the patient is on iron chelation and presents with sepsis or diarrhoea, *Yersinia Enterocolitica* or *Klebsiella* infection should be considered and can usually be treated with gentamicin IV in addition to IV ceftriaxone 2g daily. IV gentamicin plus teicoplanin 6mg/kg can be given in septic patients who are known to be penicillin allergic. Seek local microbiology advice.

Malaria is expected to be more severe in those with SCD. Specifically ask about travel to areas with endemic malaria.

In mild cases, when the patient could go home or where an early discharge is anticipated, oral antibiotics can be considered.

Acute anaemia

Individuals with symptomatic anaemia and a haemoglobin significantly lower than their usual baseline (which in sickle cell disease ranges widely e.g. from 60-110 g/L) are likely to need red cell transfusion. Ideally discuss transfusion requirements in more complex cases with a haematologist, depending upon local arrangements and policy.

An aplastic crisis may also be more readily observed in SCD as red cell lifespan is shortened, resulting in a rapid drop in haemoglobin with bone marrow dysfunction. A low reticulocyte count can predict a marked fall in haemoglobin. Urgent haematology advice is required.

Priapism

Priapism is a painful penile erection, which persists despite lack of sexual desire/activity. Patients are instructed to present to ED within two hours of onset as there is a risk of irreversible damage if fulminant priapism lasts for longer than four hours.

Management of priapism can include more conservative measures such as gentle exercise, analgesia, hydration, bladder emptying/passing urine, warmth and alpha-adrenergic agonist (e.g. etilefrine 15mg 6-hourly, maximum dose 60mg in 24 hours). Cold compresses and ice packs are contra-indicated. If these measures do not lead to rapid resolution of the priapism, the patient may require an urgent drainage procedure with phenylephrine instillation. This procedure can be carried out in the ED by a suitably trained and experienced clinician.

Exchange transfusion may be required early in resistant or recurrent cases. Review by urology should take place within three hours of presentation to avoid potential permanent complications. Haematology referral is also advised.

Abdominal pain

Liver and splenic sequestration crises can lead to rapid organ enlargement, particularly in children, and a consequential anaemia. Consider further investigations to assess for potential complications of gall stone disease and appendicitis in those presenting with ongoing abdominal pain.

Pregnancy-related presentations

There is an increased risk of pre-eclampsia and maternal and neonatal mortality in individuals with SCD. Contact Haematology and Obstetrics teams for urgent input.

Ongoing care

Where available, use day unit settings in which staff have specialist knowledge and training for assessment and treatment of patients presenting with VOE. Haematology Day Units can allow patients to be treated in an appropriate environment by staff familiar with the management of acute sickle pain.

Patients with VOE should always be cared for in an age-appropriate setting.

For pregnant women with an acute painful sickle cell episode, seek advice from the obstetrics team. From 16 weeks gestation, patients may be more appropriately admitted under obstetrics on labour ward HDU with regular specialist sickle input, although local policies will vary.

Staff training

All healthcare professionals who may care for patients with VOE should receive regular training, with topics including:

- Pain monitoring, effective analgesia, and patient-focused prescribing behaviours
- Recognition and identification of potential acute complications
- Attitudes towards and preconceptions about patients with sickle cell disease including those presenting with VOE.

All healthcare professionals in emergency departments who care for acutely unwell patients with sickle cell disease should have access to locally agreed protocols and specialist support.

Discharge considerations

Before discharge, provide the patient (and/or carer) with information on how to continue to manage the current episode, including:

- how to obtain specialist support (*signposting*)
- how to obtain additional medication (*therapeutic safety-netting*)
- how to manage any side effects of treatment e.g. opiate-associated constipation such as activity, a high fibre diet and laxative use (*clinical safeguarding*).

Once an individual is able to manage their pain on oral analgesia, is afebrile and there are no ongoing requirements for oxygen supplementation, discharge can be considered.

There should be a clear written plan for de-escalation of opiates (and counselling regarding risk of opiate dependency) outlined in the discharge summary, particularly if there has been a need for slow release opiates in addition to immediate release opiates.

If there is a requirement for ongoing opiate medication, there should be a clear written plan for where the patient can obtain future prescriptions, and a maximum of 2-week supply of opiates may be supplied from secondary care. Clear information should be shared with primary healthcare and community services for the unusual situation in which ongoing opiate prescriptions are required.

About this document

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Review

Further review usually within three years or sooner if important information becomes available.

Declaration of Interest

None declared.

Disclaimers

The College recognises that patients, their situations, Emergency Departments and staff all vary. This guideline cannot cover all possible scenarios. The ultimate responsibility for the interpretation and application of this guideline, the use of current information and a patient's overall care and wellbeing resides with the treating clinician.

Research recommendations

Evidence for formulations, adjunctive therapies and routes for effective pain management.

Non-pharmacological interventions, e.g. rehabilitative on recovery and pain management.

Effect of Same Day Emergency Care, or specialist day care unit on patient-focussed outcomes.

Audit standards

Audit of the management of acute pain in those with sickle cell disease is usually carried out annually, with a standardised proforma based on the NICE audit form. Results are discussed in HCC meetings and relevant action should be taken to address any shortcomings.

Time to analgesia audit standard – a modification of the NICE acute pain audit proforma but with time of triage and time of administration of analgesia and clear time of out of hospital analgesia including that administered by ambulance paramedics prior en-route to the Emergency Department.

Key words for search

Sickle cell disease, sickle cell disorders, sickle cell crisis, sickle cell anaemia, vaso-occlusive episode, acute pain, acute chest syndrome, emergency department.

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Appendix 1 – Example of an SCD Clinical Pathway

ED management of acute SCD presentations

Triage as high priority (level 1) and assess in a 'main' ED area

Age-appropriate pain score/ VAS; assess prior to analgesia
Record observations (Temp, HR, RR, SpO₂, BP, NEWS/PEWS, weight)

Analgesia within 15 minutes, opiates if severe pain (≥7/10)
ESCALATE PROMPTLY if NEWS ≥4, or 3 in any category, SpO₂ < 95% on air; increasing O₂ requirement

SUPPORTIVE CARE – see details

DECISION < 1hr

Uncomplicated SCD acute pain?

YES - uncomplicated acute pain

DECISION ~2 hrs

Can they go to a Day Care Unit?

- ✓ Unit available? e.g. Weekday 9-4pm
- ✓ Ambulant/wheelchair?
- ✓ O₂ Sats ≥ 94% or <10% below baseline?
- ✓ No Acute Chest Syndrome, priapism or non-sickle pathology (e.g. GI bleed)

Adults - if pain settles after 2 opiate doses & well, share decision making with patient, consider discharge after 4 hours with FU plan, opiate de-escalation & emergency contacts.
Children and pregnancy - low admission threshold

ANALGESIA WITHIN 15 MINUTES of arrival
 USE INDIVIDUAL CARE PLAN if available. For children/infants a generic care plan is often used e.g. oral morphine.
 If no care plan, as per NICE Guideline 143, give morphine sc 0.05-0.1 mg/kg/30 minutes to max 3 doses until pain controlled. then 0.05-0.1 mg/kg 2-4 hourly.

SUPPORTIVE CARE
 N.B. Patients & carers are SCD EXPERTS; consider non-sickle cause if pain atypical for sickle; empathy and respect
 Call bell; warm environment, blankets and drinking water
Oxygenation – maintain ≥95% with supplemental O₂
Anti-emetic (e.g. metoclopramide)
Oral analgesia (e.g. paracetamol; ibuprofen, codeine)
Anti-pruritic (e.g. chlorphenamine)
Hydration: aim for euvoelaemia orally; IV if required.
Observation, pain & NEWS score at 30 minutes then hourly for 1-6 hrs, then 4 hourly
Antibiotics – if fever or signs of infection N.B. hyposplenic
Thromboprophylaxis in adults, unless contraindicated
Laxatives when on opiate analgesia

NO – complicated e.g. fever ≥ 38°C; O₂ (RA) <95%, chest pain, unresolved priapism, ongoing pain despite oral analgesia and ≥ 3 opiate doses, presence of new neurological symptoms

Full history, examination and investigations
 e.g. FBC (retics), renal & liver chemistry, CRP, G&S, +/- BC, MSU, CXR, ABG/VBG, sputum MC&S, resp virus PCR
Discuss promptly with Haematologist and admitting team +/- liaise with HDU/ITU

Admission

Exclude specific SCD complications (details below)
Discuss with haematology and admitting team
 N.B. **Age >16-18* likely adult admission;**
Pregnancy > 16-23/40 likely obstetrics admission*
 Consider paediatric ward, if available, for teenagers not yet transitioned to adult services
 If ED site is separate to haematology admission site, **patient requires urgent ambulance transfer**
 *local agreements vary

Specific SCD complications: Prompt discussion with specialist teams e.g. within 30 minutes

Neurological events: new neurological symptoms need urgent investigation; ischaemic strokes and intracranial haemorrhage need urgent discussion with HASU (hyperacute stroke unit), Neurology and Haematology teams and urgent neuroimaging; management includes **urgent exchange transfusion and liaison with stroke team**

Infection/sepsis: pneumococcal & other infections; septic screen, antibiotics according to likely focus; malaria

Acute chest syndrome: new CXR pulmonary opacity, respiratory symptoms +/- fever = **LEADING CAUSE OF DEATH**
Discuss promptly with Haematologist +/- liaise with HDU/ITU as likely need for transfusion
 Differential diagnosis: CAP – oxygen, culture, antibiotics & atypical cover (e.g. IV co-amoxiclav & clarithromycin)

Acute anaemia: discuss transfusion requirements with haematologist; aplastic crisis with low reticulocytes

Priapism: painful penile erection, present ≤2 hours of onset, risk of irreversible damage if >4 hours duration
 Management: analgesia, hydration, bladder emptying, gentle exercise, oral etilefrine 15mg stat; do **not** pack in ice, unresolving priapism requires aspiration +/- exchange transfusion; urgent urology & haematological review

Abdominal pain: liver & splenic sequestration, esp. children; mesenteric; complications of gall stone disease,

Pregnancy: increased pre-eclampsia, maternal and neonatal mortality; **contact Haematology and Obstetrics teams**

Appendix 2 – The ACT NOW* paradigm

A ANALGESIA	Give analgesia within 30 minutes [NICE guidance] Regular assessment of pain score every 30 minutes until controlled and then every 4 hours. Refer to individualised care plan.
C OMPASSION	Compassion , kind, actively listen, provide reassurance and keep informed Consider IV fluids & antibiotics to treat infection and dehydration.
T EST/TRIGGER	Tests: transfusion history/previous transfusion reactions. Blood tests (FBC, reticulocyte count, group and save, routine renal, liver & bone biochemistry, CRP), other tests as suggested by history e.g. CXR, MSU. Trigger – what precipitated the crisis? e.g. infection, dehydration, hypoxia, travel, pregnancy, stress, cold exposure.
N OTIFY	Notify Specialist Haematology Team Notify Next of Kin or advocate (when requested or in individual care plan)
O XYGEN	Offer oxygen supplementation if saturations <95% RA; regularly monitor oxygen saturations, including on room air, hourly for first 6 hours and then 4 hourly if stable as per NICE guidance
W ATCH	Watch and keep warm – regular observations of BP, pulse, respiratory rate, SpO ₂ , temperature. Assess pain every 30 minutes until controlled. Escalate promptly (use local scoring e.g. NEWS2 for adults). Encourage fluids.

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*ACT NOW acronym developed by the Pan London Sickle Cell Steering group involving NHSE, Public Health Clinicians, networks, clinicians and patient/ carer representatives.



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