

Sickle cell disease: managing acute painful episodes in hospital

Clinical guideline

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Your responsibility

The recommendations in this guideline represent the view of NICE, arrived at after careful consideration of the evidence available. When exercising their judgement, professionals and practitioners are expected to take this guideline fully into account, alongside the individual needs, preferences and values of their patients or the people using their service. It is not mandatory to apply the recommendations, and the guideline does not override the responsibility to make decisions appropriate to the circumstances of the individual, in consultation with them and their families and carers or guardian.

All problems (adverse events) related to a medicine or medical device used for treatment or in a procedure should be reported to the Medicines and Healthcare products Regulatory Agency using the [Yellow Card Scheme](#).

Local commissioners and providers of healthcare have a responsibility to enable the guideline to be applied when individual professionals and people using services wish to use it. They should do so in the context of local and national priorities for funding and developing services, and in light of their duties to have due regard to the need to eliminate unlawful discrimination, to advance equality of opportunity and to reduce health inequalities. Nothing in this guideline should be interpreted in a way that would be inconsistent with complying with those duties.

Commissioners and providers have a responsibility to promote an environmentally sustainable health and care system and should [assess and reduce the environmental impact of implementing NICE recommendations wherever possible](#).

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This guideline is the basis of QS58.

Overview

This guideline covers managing acute painful sickle cell episodes in children, young people and adults who present at hospital, from presentation until when they are discharged. It aims to reduce variation in how acute episodes are managed in hospital, focusing on effective, prompt and safe pain relief.

Who is it for?

- Healthcare professionals
- People with sickle cell disease and their families and carers

Introduction

Sickle cell disease is the name given to a group of lifelong inherited conditions of haemoglobin formation. Most people affected are of African or African-Caribbean origin, although the sickle gene is found in all ethnic groups. Sickle cell disease can have a significant impact on morbidity and mortality.

It is estimated that there are between 12,500 and 15,000 people with sickle cell disease in the UK. The prevalence of the disease is increasing because of immigration into the UK and new births. The NHS Sickle Cell and Thalassaemia Screening Programme also means that more cases are being diagnosed.

Acute painful sickle cell episodes (also known as painful crises) are caused by blockage of the small blood vessels. The red blood cells in people with sickle cell disease behave differently under a variety of conditions, including dehydration, low oxygen levels and elevated temperature. Changes in any of these conditions may cause the cells to block small blood vessels and cause tissue infarction. Repeated episodes may result in organ damage.

Acute painful sickle cell episodes occur unpredictably, often without clear precipitating factors. Their frequency may vary from less than 1 episode a year to severe pain at least once a week. Pain can fluctuate in both intensity and duration, and may be excruciating. Most of the painful episodes are managed at home, with patients usually seeking hospital care only if the pain is uncontrolled or they have no access to analgesia. Patients who require admission may remain in hospital for several days. The primary goal in the management of an acute painful sickle cell episode is to achieve effective pain control both promptly and safely.

The management of acute painful sickle cell episodes for patients presenting at hospital is variable throughout the UK, and this is a frequent source of complaints from patients. Common problems include unacceptable delays in receiving analgesia, insufficient or excessive doses, inappropriate analgesia, and stigmatising the patient as drug seeking.

This guideline addresses the management of an acute painful sickle cell episode in patients presenting to hospital until discharge. This includes the use of pharmacological and non-pharmacological interventions, identifying the signs and symptoms of acute complications, skills and settings for managing an acute painful episode, and the

information and support needs of patients.

This is an overarching guideline covering the principles of how to manage an acute painful sickle cell episode in hospital. Local protocols should be referred to for specific management plans, including drug choice and dosages. This guideline includes the management of acute painful sickle cell episodes in children and young people and in pregnant women. The guideline recommendations apply to all patients presenting with an acute painful sickle episode unless there are differences in management for these groups, in which case these are clearly outlined.

Recommendations

People have the right to be involved in discussions and make informed decisions about their care, as described in [NICE's information on making decisions about your care](#).

[Making decisions using NICE guidelines](#) explains how we use words to show the strength (or certainty) of our recommendations, and has information about prescribing medicines (including off-label use), professional guidelines, standards and laws (including on consent and mental capacity), and safeguarding.

Individualised assessment at presentation

[NICE has produced a guideline on babies, children and young people's experience of healthcare.](#)

- 1.1.1 Treat an acute painful sickle cell episode as an acute medical emergency. Follow locally agreed protocols for managing acute painful sickle cell episodes and/or acute medical emergencies that are consistent with this guideline.
- 1.1.2 Throughout an acute painful sickle cell episode, regard the patient (and/or their carer) as an expert in their condition, listen to their views and discuss with them:
 - the planned treatment regimen for the episode
 - treatment received during previous episodes
 - any concerns they may have about the current episode
 - any psychological and/or social support they may need.
- 1.1.3 Assess pain and use an age-appropriate pain scoring tool for all patients presenting at hospital with an acute painful sickle cell episode.
- 1.1.4 Offer analgesia within 30 minutes of presentation to all patients presenting at

hospital with an acute painful sickle cell episode (see also the [recommendations on primary analgesia](#)).

1.1.5 Clinically assess all patients presenting at hospital with an acute painful sickle cell episode, including monitoring of:

- blood pressure
- oxygen saturation on air (if oxygen saturation is 95% or below, offer oxygen therapy)
- pulse rate
- respiratory rate
- temperature.

Be aware that some pulse oximeters can underestimate or overestimate oxygen saturation levels, especially if the saturation level is borderline.

Overestimation has been reported in people with dark skin. See also the [NHS England Patient Safety Alert on the risk of harm from inappropriate placement of pulse oximeter probes](#).

1.1.6 Assess all patients with sickle cell disease who present with acute pain to determine whether their pain is being caused by an acute painful sickle cell episode or whether an alternative diagnosis is possible, particularly if pain is reported as atypical by the patient.

Primary analgesia

1.1.7 When offering analgesia for an acute painful sickle cell episode:

- ask about and take into account any analgesia taken by the patient for the current episode before presentation
- ensure that the drug, dose and administration route are suitable for the severity of the pain and the age of the patient

- refer to the patient's individual care plan if available.
- 1.1.8 Offer a bolus dose of a strong opioid by a suitable administration route, in accordance with locally agreed protocols for managing acute painful sickle cell episodes, to:
- all patients presenting with severe pain
 - all patients presenting with moderate pain who have already had some analgesia before presentation.
- 1.1.9 Consider a weak opioid as an alternative to a strong opioid for patients presenting with moderate pain who have not yet had any analgesia.
- 1.1.10 Offer all patients regular paracetamol and NSAIDs (non-steroidal anti-inflammatory drugs) by a suitable administration route, in addition to an opioid, unless contraindicated.
- The use of NSAIDs should be avoided during pregnancy, unless the potential benefits outweigh the risks. NSAIDs should be avoided for treating an acute painful sickle cell episode in women in the third trimester. See the BNF for details of contraindications.
- 1.1.11 Do not offer pethidine for treating pain in an acute painful sickle cell episode.

Reassessment and ongoing management

- 1.1.12 Assess the effectiveness of pain relief:
- every 30 minutes until satisfactory pain relief has been achieved, and at least every 4 hours thereafter
 - using an age-appropriate pain scoring tool
 - by asking questions, such as:
 - How well did that last painkiller work?

- Do you feel that you need more pain relief?
- 1.1.13 If the patient has severe pain on reassessment, offer a second bolus dose of a strong opioid (or a first bolus dose if they have not yet received a strong opioid).
- 1.1.14 Consider patient-controlled analgesia if repeated bolus doses of a strong opioid are needed within 2 hours. Ensure that patient-controlled analgesia is used in accordance with locally agreed protocols for managing acute painful sickle cell episodes and/or acute medical emergencies.
- 1.1.15 Offer all patients who are taking an opioid:
- laxatives on a regular basis
 - anti-emetics as needed
 - antipruritics as needed.
- 1.1.16 Monitor patients taking strong opioids for adverse events, and perform a clinical assessment (including sedation score):
- every 1 hour for the first 6 hours
 - at least every 4 hours thereafter.
- 1.1.17 If the patient does not respond to standard treatment for an acute painful sickle cell episode, reassess them for the possibility of an alternative diagnosis.
- 1.1.18 As the acute painful sickle cell episode resolves, follow locally agreed protocols for managing acute painful sickle cell episodes to step down pharmacological treatment, in consultation with the patient.

Possible acute complications

- 1.1.19 Be aware of the possibility of acute chest syndrome in patients with an acute painful sickle cell episode if any of the following are present at any time from presentation to discharge:

- abnormal respiratory signs and/or symptoms
- chest pain
- fever
- signs and symptoms of hypoxia:
 - oxygen saturation of 95% or below **or**
 - an escalating oxygen requirement.

Be aware that some pulse oximeters can underestimate or overestimate oxygen saturation levels, especially if the saturation level is borderline. Overestimation has been reported in people with dark skin. See also the [NHS England Patient Safety Alert on the risk of harm from inappropriate placement of pulse oximeter probes.](#)

1.1.20 Be aware of other possible complications seen with an acute painful sickle cell episode, at any time from presentation to discharge, including:

- acute stroke
- aplastic crisis
- infections
- osteomyelitis
- splenic sequestration.

Management of underlying pathology

1.1.21 Do not use corticosteroids in the management of an uncomplicated acute painful sickle cell episode.

Non-pharmacological interventions

1.1.22 Encourage the patient to use their own coping mechanisms (for example, relaxation techniques) for dealing with acute pain.

Settings and training

1.1.23 All healthcare professionals who care for patients with an acute painful sickle cell episode should receive regular training, with topics including:

- pain monitoring and relief
- the ability to identify potential acute complications
- attitudes towards and preconceptions about patients presenting with an acute painful sickle cell episode.

1.1.24 Where available, use day care settings in which staff have specialist knowledge and training for the initial assessment and treatment of patients presenting with an acute painful sickle cell episode.

1.1.25 All healthcare professionals in emergency departments who care for patients with an acute painful sickle cell episode should have access to locally agreed protocols and specialist support from designated centres.

1.1.26 Patients with an acute painful sickle cell episode should be cared for in an age-appropriate setting.

1.1.27 For pregnant women with an acute painful sickle cell episode, seek advice from the obstetrics team and refer when indicated.

Discharge information

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

- how to obtain specialist support
- how to obtain additional medication
- how to manage any potential side effects of the treatment they have received in hospital.

Terms used in this guideline

Moderate pain

Pain with a Visual Analogue Scale (VAS; or equivalent) score typically within the range of 4 to 7 (this description should not be interpreted as a strict definition and will not apply to all patients, as pain is subjective).

Patient-controlled analgesia (PCA)

A method of safely administering strong opioids which is controlled by the patient (or a nurse for nurse-controlled analgesia).

Severe pain

Pain with a VAS (or equivalent) score typically above 7 (this description should not be interpreted as a strict definition and will not apply to all patients, as pain is subjective).

Recommendations for research

The guideline development group has made the following recommendations for research.

1 Pain management for patients with an acute painful sickle cell episode

For patients with an acute painful sickle cell episode, what are the effects of different opioid formulations, adjunct pain therapies and routes of administration on pain relief and acute sickle cell complications?

Why this is important

Limited evidence is available on the effectiveness of different opioid formulations, routes of administration and adjunct therapies in the treatment of an acute painful sickle cell episode. A series of randomised controlled trials (RCTs) should be conducted that compare the effects of different opioid formulations, adjunct pain therapies and routes of administration. These RCTs should be conducted separately in adults and children, and cover the duration of the acute painful episode. Outcomes should include pain and adverse events such as acute chest syndrome.

2 Use of low-molecular-weight heparin to treat patients with an acute painful sickle cell episode

Are therapeutic doses of low-molecular-weight heparin (LMWH) effective, compared with prophylactic doses of LMWH, in reducing the length of stay in hospital of patients with an acute painful sickle cell episode?

Why this is important

Moderate-quality evidence from one RCT suggested a significant benefit of treating patients with an acute painful sickle cell episode with LMWH. This was supported by exploratory health economic analyses suggesting a large reduction in length of stay and associated costs. An RCT should be conducted that examines the effect of therapeutic

doses of LMWH, compared with prophylactic doses, on the length of stay in hospital of patients with an acute painful sickle cell episode. The RCT should be conducted separately in adults and children, and cover the duration of the painful episode.

3 Non-pharmacological interventions for patients with an acute painful sickle cell episode

For patients with an acute painful sickle cell episode, are non-pharmacological interventions, such as massage, effective in improving their recovery from the episode?

Why this is important

There was a lack of evidence on the potential benefits of supportive interventions for patients with an acute painful sickle cell episode. An RCT should be conducted that examines the effect of providing rehabilitation interventions that are aimed at improving a patient's recovery after an acute painful sickle cell episode. Such interventions could include massage and physical therapy. The intervention should be provided within the hospital setting, and patients should be followed up 7 days after the episode. Data should be collected to inform outcomes such as length of stay, health-related quality of life and coping strategies.

4 Cost effectiveness of daycare units for treating patients with an acute painful sickle cell episode

Are day care units cost effective compared with emergency settings for treating patients with an acute painful sickle cell episode?

Why this is important

There was a lack of evidence on the cost effectiveness of day care units for treating patients with an acute painful sickle cell episode in the UK. A trial should be carried out that compares treating patients with an acute painful sickle cell episode in an emergency department setting and in a specialist sickle cell day care unit. Outcomes should include health-related quality of life (HRQoL). Data should be collected using validated measure(s) of HRQoL, including EQ-5D.

Finding more information and committee details

To find NICE guidance on related topics, including guidance in development, see the [NICE topic page on blood conditions](#).

For full details of the evidence and the guideline committee's discussions, see the [full guideline](#). You can also find information about [how the guideline was developed](#), including [details of the committee](#).

NICE has produced [tools and resources to help you put this guideline into practice](#). For general help and advice on putting our guidelines into practice, see [resources to help you put NICE guidance into practice](#).

Update information

Minor changes since publication

October 2022: We added text to indicate that pulse oximetry may be less reliable in people with dark skin. We also added a link to the NHS patient safety alert on the risk of harm from inappropriate placement of pulse oximeter probes. See recommendations 1.1.5 and 1.1.19.

November 2021: We added a link to NICE's guideline on babies, children and young people's experience of healthcare in section 1.

August 2016: We deleted an out-of-date research recommendation.

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