NATIONAL INSTITUTE FOR HEALTH AND CARE EXCELLENCE

Health and social care directorate

Quality standards and indicators

Briefing paper

Quality standard topic: Sickle cell crisis

Output: Prioritised quality improvement areas for development.

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Contents

1 Introduction

This briefing paper presents a structured overview of potential quality improvement areas for sickle cell crisis. It provides the Committee with a basis for discussing and prioritising quality improvement areas for development into draft quality statements and measures for public consultation.

1.1 Structure

This briefing paper includes a brief description of the topic, a summary of each of the suggested quality improvement areas and supporting information.

If relevant, recommendations selected from the key development source below are included to help the Committee in considering potential statements and measures.

1.2 Development source

The key development source(s) referenced in this briefing paper is:

- Sickle cell acute painful episode. NICE clinical guideline 143 (2012).

2 Overview

2.1 Focus of quality standard

This quality standard will cover the management of a sickle cell crisis in people presenting to hospital until discharge.

2.2 Definition

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.

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 one episode a year to

severe pain at least once a week. Pain can fluctuate in both intensity and duration, and may be excruciating.

2.3 Incidence and prevalence

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.

Sickle cell disease can have a significant impact on morbidity and mortality.

2.4 Management

The majority of 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.

2.5 Peer review programmes

Services for children and young people with haemoglobin disorders and adults with haemoglobin disorders were the subject of a peer review programme lead by the West Midlands Quality Review Service and supported by the UK forum on Haemoglobin Disorders and the NHS Sickle Cell and Thalassaemia Screening Programme. The purpose of the reviews was to improve the quality of services for people with haemoglobin disorders.

The peer review programme for services for children and young people with haemoglobin disorders took place in 2010/11. 16 hospitals in England delivering specialist services and three large linked hospitals were reviewed. The peer review programme for services for adults with haemoglobin disorders took place in 2012/13. 29 hospitals covering 32 teams delivering services for adults were visited. Where applicable the results of the audit have been included in the current practice sections of the briefing paper.

2.6 National Outcome Frameworks

Table 1 show the outcomes, overarching indicators and improvement areas from the framework that the quality standard could contribute to achieving.

Table 1 NHS Outcomes Framework 2013/14

Domain	Overarching indicators and improvement areas
1 Preventing people from dying prematurely	***Overarching indicator*** 1a Potential years of life lost (PYLL) from causes considered amenable to healthcare i adults ii *children and young people (placeholder)*
2 Enhancing quality of life for people with long-term conditions	***Overarching indicator*** 2 Health-related quality of life for people with long-term conditions** ***Improvement areas*** **Ensuring people feel supported to manage their condition** 2.1 Proportion of people feeling supported to manage their condition**
4 Ensuring that people have a positive experience of care	***Overarching indicator*** 4b Patient experience of hospital care ***Improvement areas*** **Improving people's experience of accident and emergency services** 4.3 Patient experience of A&E services ***Improving children and young people's experience of healthcare (placeholder)***
Alignment across the health and social care system ** Indicator complementary with Adult Social Care Outcomes Framework (ASCOF)	

3 Summary of suggestions

3.1 Responses

In total 11 stakeholders responded to the 2-week engagement 17/07/2013 – 31/07/2013, this includes 3 stakeholders who responded but did not suggest any areas for quality improvement.

Stakeholders were asked to suggest up to 5 areas for quality improvement. Specialist committee members were also invited to provide suggestions. The responses have been merged and summarised in table 2 for further consideration by the Committee.

Full details on the suggestions provided are given in appendix 1 for information.

Table 2 Summary of suggested quality improvement areas

Suggested area for improvement	Stakeholders
Assessment at presentation • Assessment • Time to delivery	BPS, Napp, NHS England, SCM, SCS, UKFHD
Primary analgesia	Napp, SCM, SCS, UKFHD
Reassessment	NHS England, SCM, RCPath & BCSH
Ongoing management • Ongoing pain relief • Non-pharmacological interventions	SCM, BPS
Acute complications	SCM, UKFHD
Settings and training • Training • Daycare settings • Access to specialised care • Designated clinical areas • Home support	NHS England, Napp, PSF, RCPath & BCSH, SCM, SCS, UKFHD
Discharge and follow up • Discharge • Follow up	BPS, Nordic, SCM
Additional areas • Awareness campaigns • Research and clinical trials • Transfusions • Management of adolescent males	NHS England, SCM, SCS, UKFHD

BPS, British Pain Society;

Napp, Napp Pharmaceuticals Limited;

NHSE, NHS England;

Nordic, Nordic Pharma Ltd;

RCPath & BCSH; Royal College of Pathologists and British Committee for Standards in Haematology;

SCS, Sickle Cell Society;

UKFHD, UK Forum on Haemoglobin Disorders;

PSF, National Health Service Commissioning Board Patient Safety Function;

SCM, Specialist Committee Member.

4 Suggested improvement areas

4.1 Assessment at presentation

4.1.1 Summary of suggestions

Assessment

Pain should be assessed promptly at presentation. If acute pain is not recognised promptly it may lead to unnecessary suffering; inappropriate discharge from A&E; inappropriate initiation of inadequate medication and inappropriate initiation of invasive medication when it is not needed.

A thorough assessment at presentation is needed to ensure people in sickle cell crisis are accurately diagnosed and people who present with non-sickle cell disease pain are not mis-diagnosed with sickle cell crisis.

Pain scores need to be undertaken as part of the assessment using a validated, age appropriate pain tool to ensure adequate analgesia upon presentation and for ongoing management. Where initial assessment of pain is undertaken, stakeholders report there is not always documentation of a validated tool being used. Consideration needs to be given to the most appropriate pain tools for children, particularly the very young or those with a cognitive impairment. Stakeholders report there is evidence for variable management of acute pain, with some underprescribing / delays and some overprescribing with adverse outcomes including opioid toxicity resulting in respiratory depression.

Time to delivery

People presenting with sickle cell disease pain need to have fast access to pain medication which will have a quick effect on their pain. Delays in analgesia can cause distress to people in sickle cell crisis and has a big impact on patient experience. Lack of adequate, rapid pain relief can lead to a breakdown in the relationship between the patient and care provider and in the long term may lead to patients avoiding coming into hospital because of previous experiences of long waiting times, damaging their health. In addition to poor patient experience, prolonged pain was reported to cause bone and tissue damage. If sickle cell disease pain is not treated promptly and expertly then there is a risk of pain escalating and necessitating admission for more invasive management.

4.1.2 Selected recommendations from development source

Table 3 below highlights recommendations that have been provisionally selected from the development source(s) that may support potential statement development. These are presented in full to inform the Committee's discussion.

Table 3 Specific areas for quality improvement

Suggested quality improvement area	Source guidance recommendations
Assessment	NICE CG143 Recommendations 1.1.1, 1.1.2, 1.1.3, 1.1.5, 1.1.6.
Time to delivery of analgesia	NICE CG143 Recommendation 1.1.4

Assessment

NICE CG143 Recommendation number 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.

NICE CG143 Recommendation number 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.

NICE CG143 Recommendation number 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.

NICE CG143 Recommendation number 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.

NICE CG143 Recommendation number 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.

Time to delivery of analgesia

NICE CG143 Recommendation number 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 recommendations 1.1.7 to 1.1.11).

4.1.3 Current UK practice

Assessment

The National Confidential Enquiry into Patient Outcomes and Death (NCEPOD) 2008, A sickle crisis?[1] reviewed the circumstances around the deaths in people with haemoglobinopathies through case note review and questionnaires. The report found assessment of pain, sedation and respiratory rate was infrequently performed. Through casenote review of the 35 people who died in hospital, 19 had pain as the admitting complaint and in only six patients was there evidence in the casenotes that pain was formally assessed using a pain assessment chart.

Time to delivery

Stakeholders reported from personal experience, having recently had several admissions to hospital and accompanying others for admission that the 30 minute guideline recommendation for pain relief had not been met on occasions.

The peer review programme of services for children and young people with haemoglobin disorders reported that several services had made efforts to make care responsive to the needs of children and young people with haemoglobin disorders

[1] A Sickle Crisis? A report of the National Confidential Enquiry into Patient Outcome and Death (2008)

however some problems were common including delays in administration of analgesia for children presenting with acute pain crisis[2]. The peer review programme recommended that all trusts who provide urgent care for children with haemoglobin disorders should audit time from presentation to first analgesia, against a standard of 30 minutes.

The peer review programme of services for adults with haemoglobin disorders[3] reported that pain management had been audited in most teams and results were inconsistent ranging from 100% to 13% of adults receiving analgesia within 30 minutes of presentation with acute pain. This was reflected in the feedback from the users who reported unacceptable delays in receiving analgesia in some emergency department and a perception of a lack of empathy amongst medical and nursing staff in these areas.

The NCEPOD 2008, A sickle crisis?[4] report found, through casenote review that of the 35 people who died in hospital, 19 had pain as the admitting complaint. The report had hoped to determine the time from admission to the first analgesia but due to poor documentation this was only possible to verify in two patients. Both of these patients received their first analgesia in the emergency department 90 minutes after admission.

[2] Service for children and young people with haemoglobin disorders peer review programme (2010-11) Overview report.
[3] Service for adults with haemoglobin disorders peer review programme (2012-13) Overview report
[4] A Sickle Crisis? A report of the National Confidential Enquiry into Patient Outcome and Death (2008)

4.2 Primary analgesia

4.2.1 Summary of suggestions

People presenting with sickle cell crisis need to have fast access to pain medication which will have a quick effect on their pain. Ineffective pain relief can cause distress to patients and is often a source of complaints and frustration from people with sickle cell and their families. Stakeholders reported there was evidence of variable management with some under prescribing / delays and some over prescribing of medication. It was felt that since sickle cell is a long term condition and because of the episodic nature of crises, the variability in the quality of care is a serious cause of concern.

Stakeholders reported oral medication offers a fast and easy solution and allow staff the opportunity to assess progress before initiating PCA medications which requires the availability of equipment and staff trained in its use.

Stakeholder highlighted the lack of choice of analgesia for children, which is limited to a small number of drugs.

4.2.2 Selected recommendations from development source

Table 4 below highlights recommendations that have been provisionally selected from the development source(s) that may support potential statement development. These are presented in full to inform the Committee's discussion.

Table 4 Specific areas for quality improvement

Suggested quality improvement area	Source guidance recommendations
Primary analgesia	NICE CG143 Recommendations 1.1.7, 1.1.8, 1.1.9, 1.1.10 and 1.1.11.

NICE CG143 Recommendation number 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.

NICE CG143 Recommendation number 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.

NICE CG143 Recommendation number 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.

NICE CG143 Recommendation number 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[1].

NICE CG143 Recommendation number 1.1.11

Do not offer pethidine for treating pain in an acute painful sickle cell episode.

4.2.3 Current UK practice

The NCEPOD 2008, A sickle crisis?[5] reported a recurring theme from the study was the excessive doses of opioid analgesics in patients with painful crisis. In nine of the 35 patients it was the advisors opinion that excessive doses of opioids had been given and of these there were five patients in whom complications occurred due to opioid overdose which contributed to the ultimate death of these patients.

[5] A Sickle Crisis? A report of the National Confidential Enquiry into Patient Outcome and Death (2008)

4.3 Reassessment

4.3.1 Summary of suggestions

Stakeholders highlighted it was essential that people in sickle cell crisis should have ongoing assessment to assess the adequacy of analgesia received at admission and to ensure repeated doses of analgesia once pain relief wore off and pain returned. Analgesia administration guidelines are based upon pain scores. To ensure adequate analgesia for ongoing management, pain scores need to be undertaken using a validated, age appropriate pain tool. Stakeholders suggested regular observations at least of pain score, respiratory rate and sedation score on an hourly basis during first six hours of attendance in emergency department with acute pain were needed.

4.3.2 Selected recommendations from development source

Table 5 below highlights recommendations that have been provisionally selected from the development source(s) that may support potential statement development. These are presented in full to inform the Committee's discussion.

Table 5 Specific areas for quality improvement

Suggested quality improvement area	Source guidance recommendations
Reassessment	NICE CG143 Recommendation 1.1.12

NICE CG143 Recommendation number 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?

4.3.3 Current UK practice

Stakeholders reported ongoing pain assessment is often inadequate and not performed often enough or according to medical instruction. Stakeholders

highlighted that patient feedback gives evidence that reassessment is not done adequately.

The peer review programme of services for children and young people with haemoglobin disorders recommended that all trusts who provide urgent care for children with haemoglobin disorders should audit the adequacy of analgesia after one and two hours[6].

Stakeholders involved in the peer review programme of services for adults with haemoglobin disorders reported large discrepencies in rates in frequency of re-assessment of pain between different units. This was not audited as frequently as 'time to analgesia, but where it had been few units achieved this target.

The NCEPOD 2008, A sickle crisis?[7] reported assessment of pain, sedation and respiratory rate was infrequently performed. Through casenote review of the 35 people who died in hospital, 19 had pain as the admitting complaint. The advisors reviewing the casenotes judged that the frequency of pain assessments was inadequate based on the severity of pain in five of the six patients whose pain was assessed with a pain score.

[6] Service for children and young people with haemoglobin disorders peer review programme (2010-11) Overview report.
[7] A Sickle Crisis? A report of the National Confidential Enquiry into Patient Outcome and Death (2008)

4.4 Ongoing management

4.4.1 Summary of suggestions

Ongoing pain relief

A lack of pain relief can cause distress to people in sickle cell crisis and may prolong admission to hospital. Patient controlled analgesia (PCA) is an effective way of providing pain relief and should be available for all patients if they wish to use it. Stakeholders report there is clear inequity of care with some units being able to offer this service, but many others not being able to as they do not have the resources to meet the needs for ongoing analgesia in a timely manner. PCA not only enables the patient to take control of their analgesia and empowers them, it is also cost-effective.

Non-pharmacological interventions

Over-reliance on some drug management may be detrimental to some people in sickle cell crisis, especially those with underlying psychological disorders such as anxiety and depression. The use of self-management psychological interventions may be a suitable alternative.

4.4.2 Selected recommendations from development source

Table 6 below highlights recommendations that have been provisionally selected from the development source(s) that may support potential statement development. These are presented in full to inform the Committee's discussion.

Table 6 Specific areas for quality improvement

Suggested quality improvement area	Source guidance recommendations
Ongoing pain relief	NICE CG143 Recommendations 1.1.13 and 1.1.14
Non-pharmacological interventions	NICE CG143 Recommendation 1.1.22

Ongoing pain relief

NICE CG143 Recommendation number 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).

NICE CG143 Recommendation number 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.

Non-pharmacological interventions

NICE CG143 Recommendation number 1.1.22

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

4.4.3 Current UK practice

Ongoing pain relief

Stakeholders report that despite good evidence that PCAs are effective they are still not available for people with sickle cell disease in many hospitals around the country. This leads to cases of inadequate pain relief being provided and is detrimental to patient care. Even when PCAs are available they are often not available at all times of the day/week or in all hospital wards.

The peer review programme of services for adults with haemoglobin disorders[8] highlighted that depending on where patients were accommodated affected the availability of PCA. PCA was not available in all teams and availability was sometimes restricted to particular wards or times of day.

Non-pharmacological interventions

No published reports relating to current practice were highlighted by stakeholders for this quality improvement area; this area is based on stakeholder's knowledge and experience.

[8] Service for adults with haemoglobin disorders peer review programme (2012-13) Overview report

4.5 Acute complications

4.5.1 Summary of suggestions

People presenting with apparently uncomplicated crisis may have serious complications underlying the initial presentation or can develop a range of additional problems / sepsis / organ failure during admission. Careful vigilance with accurate, sufficiently frequent physiological observations forms the basis for identifying deterioration and escalating to appropriate clinical staff to allow necessary additional interventions. Failure to record complications may lead to delays in recognising over sedation with opiate drugs and development of ACS, it may be fatal if left untreated.

4.5.2 Selected recommendations from development source

Table 7 below highlights recommendations that have been provisionally selected from the development source(s) that may support potential statement development. These are presented in full to inform the Committee's discussion.

Table 7 Specific areas for quality improvement

Suggested quality improvement area	Source guidance recommendations
Acute complications	NICE CG143 Recommendations 1.1.19 and 1.1.20

NICE CG143 - Recommendation number 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.

NICE CG143 - Recommendation number 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.

4.5.3 Current UK practice

Stakeholders highlighted there are many cases – reported through adverse event recording on National Haemoglobinopathy Registry, presented at UK Forum for Haemoglobin Disorders scientific meetings, and from hospital Mortality and Morbidity reviews – of 'failure to rescue' or to adequately recognise how seriously ill patients are / become while in hospital, leading to adverse outcomes. This is in acute medical patients in general, and relevantly in those with sickle cell crisis / additional complications also.

The NCEPOD 2008, A sickle crisis?[9] reported there was room for improvement in reporting and acting on abnormal observations. The report stated it was important for observations to be routinely recorded and that any side effects and signs of deterioration are looked out for.

[9] A Sickle Crisis? A report of the National Confidential Enquiry into Patient Outcome and Death (2008)

4.6 Settings and training

4.6.1 Summary of suggestions

Training

Appropriately trained healthcare professionals are essential to ensure the quality and safety of patient care, ensuring adequate pain relief is given promptly and monitored and staff are aware of the potential complications people with sickle cell disease can develop which can have a high morbidity and mortality. Stakeholders report many people suffer early deaths unnecessarily mainly due to insufficient knowledge on sickle cell. It was reported that care is inconsistent with different health professionals and hospitals and needs to take into account the needs of children and young people.

Stakeholders report from patient feedback that a major complaint of people in sickle cell crisis is that healthcare professionals do not have sufficient knowledge about sickle cell disease. Patients do not feel safe and often encounter negative attitudes from staff. It is felt this may lead to fear of going to hospital or behaviours from patients which reinforce negative perceptions by staff.

Daycare settings

Stakeholders suggested daycare settings are an effective way of providing initial assessment and treatment for sickle cell crisis. Staff in these settings have specialist knowledge and therefore patients receive rapid and appropriate analgesia and are assessed for complications. Provision of early, effective treatment may lead to improved patient experience, improved pain control and also results in decreased hospital admissions and length of stay.

Access to specialised care

Stakeholders felt there were still insufficient places of specialist care in high prevalent areas. People with a greater knowledge of sickle cell crisis need to train others in order for specialised areas of care to be established in high sickle populations.

People with sickle cell disease can present to any emergency department. In low prevalence areas particularly this may not be a very frequent occurrence and staff may not have much experience of the management of sickle cell crisis. It is therefore important that these units have access to local protocols so there are clear guidelines for immediate care and support from specialist centres.

Designated clinical areas

Stakeholders suggested designated clinical area/s which are conducive for recovery from acute pain crisis.

Home support

Most pain episodes are managed by the person with sickle cell and their carers or families at home and they self-evaluate when they can no longer manage and need hospital care. Stakeholders suggest even more severe pain crises, which have no additional or underlying complications after adequate assessment can be treated at home if appropriate analgesia is available.

4.6.2 Selected recommendations from development source

Table 8 below highlights recommendations that have been provisionally selected from the development source(s) that may support potential statement development. These are presented in full to inform the Committee's discussion.

Table 8 Specific areas for quality improvement

Suggested quality improvement area	Source guidance recommendations
Training	NICE CG143 Recommendation 1.1.23
Daycare settings	NICE CG143 Recommendation 1.1.24
Access to specialised care	NICE CG143 Recommendation 1.1.25
Designated clinical areas	Not directly covered in NICE CG143 and no recommendations are presented
Home Support	Not directly covered in NICE CG143 and no recommendations are presented

Training

NICE CG143 - Recommendation number 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.

Daycare settings

NICE CG143 - Recommendation number 1.1.24

Where available, use daycare 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.

Access to specialised care

NICE CG143 - Recommendation number 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.

Designated clinical areas

Designated clinical areas are not directly covered in NICE clinical guideline 143 and no recommendations are presented relating to the suggested area for quality improvement.

Home Support

Home support is not directly covered in NICE clinical guideline 143 and no recommendations are presented relating to the suggested area for quality improvement.

4.6.3 Current UK practice

Training

The peer review programme of services for adults with haemoglobin disorders[10] reported training of healthcare professionals was inconsistent, and was not offered regularly or repeatedly in the majority of units reviewed even when caring for large numbers of patients with sickle cell disease. Training about haemoglobin disorders in the emergency department was often problematic. One site had mandatory training for staff, which was a robust method of ensuring that all staff were up to date. Patient feedback frequently said that healthcare professionals did not have a good knowledge of sickle cell disease and its complications. 29% of sites had a training plan to ensure that all staff are developing and maintaining appropriate competencies for their role in the care of patients with haemoglobin disorders. 35% of sites have a programme of induction and training covering the care of patients with haemoglobin disorders for clinical staff in the emergency department, non-consultant medical staff and allied health professionals.

[10] Service for adults with haemoglobin disorders peer review programme (2012-13) Overview report

NCEPOD 2008, A sickle crisis?[11] highlighted that people with sickle cell disease can develop a number of complications and it is important staff caring for the patients are aware of the possible complications as patients can deteriorate very quickly. The report gave examples where inadequacy of monitoring following high doses of opioid analgesia contributed to patient death. The report found four cases in which it was judged by the advisors that advice was not sought from external experts when required. Clinicians completing the questionnaires in the report were asked if in their opinion there were issues related to the training of the medical team and junior doctors in the management of haemoglobinopathies. 12 clinicians commented on areas of concern. There was also considered to be a lack of education – including amongst Emergency Department doctors.

Daycare settings

NICE Costing Report[12] on Sickle Cell crisis states there are a small number of daycare units in England containing specialist trained sickle cell staff. These units are generally intended to be used by patients with an uncomplicated acute painful episode during the day instead of formally admitting them to hospital.

The peer review programme of services for adults with haemoglobin disorders[13] and stakeholders involved in the programme reported that day unit facilities were not available in many units in England, even those with large sickle cell populations. Even where day units are available, opening hours are often restricted to working hours and patients were admitted out of hours. Day unit services were highly rated by patients and had high levels of patient satisfaction.

Access to specialised care

The peer review programme of services for adults with haemoglobin disorders[14] and stakeholders involved in the programme reported that emergency departments in high prevalence areas did not always have locally agreed protocols for the care of acute painful crisis and sometimes although protocols may have existed, staff were not aware of them leading to delays in patients receiving adequate analgesia. As part of the peer review programme the majority of centres in low prevalence areas were not reviewed but patient feedback would suggest that many hospitals do not have local treatment protocols and that this leads to significant delays in care. Emergency departments in low prevalence areas were not always aware of where to get specialist support from, especially out of hours. Furthermore specialist centres were not sure which hospitals they should be covering and did not provide a 24/7 service for advice. Although the 32 teams reviewed during the 29 visits were thought

[11] A Sickle Crisis? A report of the National Confidential Enquiry into Patient Outcome and Death (2008)
[12] NICE costing report (2012)
[13] Service for adults with haemoglobin disorders peer review programme (2012-13) Overview report
[14] Service for adults with haemoglobin disorders peer review programme (2012-13) Overview report

to care for the majority of adults with haemoglobin disorders in England, an unknown number of adults are cared for by haematology teams in hospitals which did not link to a specialist team or are not part of an established network of care. Several acute trusts reviewed did not link formally or informally into any network or with any specialist team. The report also highlighted that although acute complications is a key part of the haemoglobinopathy service, 28% of departments did not have clear emergency department guidelines. Even where available they were sometimes unclear and emergency department staff were not always aware of their existence.

NCEPOD 2008, A sickle crisis?[15] report showed that non Sickle causes of pain or rarer complications of Sickle may not be recognised. The organisation of care into local and responsible specialist units via CRG specification means that all should have a clear access to protocols and a route to specialist expertise when needed, particularly relevant to low prevalence areas. Three of the 26 sites reviewed had no guidelines or protocols. Less than a third (8/26) of sites had agreed policies with other hospitals for the transfer of sickle cell patients.

Designated clinical areas

No published reports relating to current practice were highlighted by stakeholders for this quality improvement area; this area is based on stakeholder's knowledge and experience.

Home support

Stakeholders reported that patients, especially those with frequent pain crises, regularly express the wish to be managed at home where possible, where they feel they recover more quickly and without the disturbance and exposure to other risks that hospital care engenders.

[15] A Sickle Crisis? A report of the National Confidential Enquiry into Patient Outcome and Death (2008)

4.7 Discharge and follow up

4.7.1 Summary of suggestions

Discharge

People in sickle cell crisis can often be discharged home whilst still in crisis. Stakeholders suggested people in sickle cell crisis and their carers needed to be fully informed on how to continue to effectively manage their pain after discharge whilst at home and of any potential symptoms that may be cause for readmission to hospital. Further information was suggested including signposting to material on pain analgesia, sexual health, psychosocial support, pregnancy and screening.

Stakeholders also suggested presentation to hospital can be due to the person experiencing other issues, for example housing issues and yet the social, psychological and wellbeing of the person is rarely considered before discharge.

Follow up

Stakeholders suggested follow up after discharge was important for several reasons; to ensure continuity of care between secondary and primary care through discharge letters, for ongoing treatment and counselling by staff at a specialist sickle cell centre which may reduce the incidence of further episodes and admissions to hospital and to reduce the risk of drug dependency as people with sickle cell may be discharged with opioid medication.

4.7.2 Selected recommendations from development source

Table 9 below highlights recommendations that have been provisionally selected from the development source(s) that may support potential statement development. These are presented in full to inform the Committee's discussion.

Table 9 Specific areas for quality improvement

Suggested quality improvement area	Source guidance recommendations
Discharge information	NICE CG143 Recommendation 1.1.28
Follow up	Not directly covered in NICE CG143 and no recommendations are presented

Discharge information

NICE CG143 - Recommendation number 1.1.19

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.

Follow up

Follow up is not directly covered in NICE clinical guideline 143 and no recommendations are presented relating to the suggested area for quality improvement.

4.7.3 Current UK practice

Discharge information

No published reports relating to current practice were highlighted by stakeholders for this quality improvement area; this area is based on stakeholder's knowledge and experience.

Follow up

No published reports relating to current practice were highlighted by stakeholders for this quality improvement area; this area is based on stakeholder's knowledge and experience

4.8 Additional areas

4.8.1 Summary of suggestions

The improvement areas below were suggested as part of the stakeholder engagement exercise however were felt to be outside the scope of the quality standard referral and the NICE guideline. There will be an opportunity for the QSAC to discuss these areas at the end of the session.

Awareness campaigns

The sickle cell population are often called ‘hard to reach’ and include people from the black and afro-caribbean communities. It is felt that people can only make an informed choice if they are given the information through more viable campaigns. These were reported to help patients and the public alike.

Research and clinical trials

Research into the effectiveness of non-opioid medication for pain and entry clinical trials were suggested by stakeholders. Research was suggested as reducing opioid medication is reported to reduce longterm side effects such as hyperalgesia, immunesuppression and hormonal changes and entry onto trials was suggested for advancing new treatments such as haematopoetic transplantation.

Transfusions

Red cell transfusions can be life-saving in sickle cell crisis, for example in aplastic crisis with profound anaemia, chest syndrome, stroke. However, they are *usually* not indicated in uncomplicated pain crisis, and always carry a degree of risk to the recipient so should be avoided if not necessary. Administration of ill-matched red cell units can lead to serious haemolytic transfusion reactions and occasionally death [for example, those which contain antigens to which the recipient has previously made an antibody reaction, even though they may no longer be detectable in current compatibility tests]. It was felt guidance was needed on the use of red cell transfusions for different presentations and complications ensuring adequate steps are taken to identify/avoid antibody formation.

Management of adolescent males

Stakeholders noted the lack of reference in the guideline to priapism which if left untreated or unrecognised may have other health implications such as impotency. Stakeholders suggested from feedback that for some adolescent males this can be a difficult subject to discuss with health professionals.

4.8.2 Selected recommendations from development source

The suggested quality improvement areas above are not directly covered in NICE CG143 and no recommendations are presented.

4.8.3 Current UK practice

Awareness campaigns

Stakeholders reported there is little to no evidence on government awareness campaigns in the UK.

No published reports relating to current practice were highlighted by stakeholders for this quality improvement area; this area is based on stakeholder's knowledge and experience.

Research and clinical trials

Stakeholders referenced that in cancer care almost all patients are entered into multicentre trials (national/international trial networks) and progress in treating various cancers is advancing fast.

No published reports relating to current practice were highlighted by stakeholders for this quality improvement area; this area is based on stakeholder's knowledge and experience.

Transfusions

Stakeholders reported there is evidence of variable transfusion practice, acutely and longer term / planned transfusion, and anecdotal reports of patients receiving unnecessary transfusion e.g. for relatively minor falls in haemoglobin, especially in hospitals less familiar with sickle cell and its complications. The risks and benefits of transfusion need to be carefully weighed in every individual case.

The 'Serious Hazards of Transfusion (SHOT)' annual report 2012[16] reported haemolytic transfusion reactions (HTR) (destruction of the donor red blood cells by host antibodies) are a significant cause of major morbidity, particularly in patients with sickle cell disease. 9 cases of HTR with major morbidity were recorded, 5 of which occurred in people with sickle cell disease. Over three years of reporting (2010–2012) there have been 16 cases of HTR in people with sickle cell disease, with 11 (68.8%) instances of major morbidity or death. Some of these reactions were potentially preventable, occurring due to failure to inform the laboratory about known sickle cell disease (so that appropriately typed red cells were not provided) and

[16] SHOT Annual Report 2012

others relate to failure in the laboratory to discover or heed previously documented alloantibodies.

The children and young people with haemoglobin disorders peer review programme[17] reported that 15 services had clinical guidelines in use covering the indicators for transfusions and arrangements for carrying out a transfusion whilst 4 services did not. The peer review visits also showed variations in the number of children with sickle cell disease on regular transfusion programmes was greater than could be explained by case mix variation and ranged from 1:10 to 1:30. The programme was not able to identify the reason for this variation.

Management of adolescent males

No published reports relating to current practice were highlighted by stakeholders for this quality improvement area; this area is based on stakeholder's knowledge and experience.

[17] Service for children and young people with haemoglobin disorders peer review programme 2010-11, Overview report.

Appendix 1: Suggestions from stakeholder engagement exercise

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
001	SCM1	Time to delivery of Analgesia in acute sickle crisis in A&E departments	Delays in analgesia cause distress to patients. Door to analgesia time of 30 mins is recommended in NICE guideline (1.1.4)	Patient feedback and audit data from recent adult peer review programme indicates marked variety across England with many patients reporting significant delays. Audit data from trusts shows many are failing to meet this target.	Adult peer review overview report due for publication soon Individual trust reports publically available on WMQRS website
002	SCM1	Analgesia appropriate for severity of pain	Ineffective pain relief causing distress to patient. (1.1.7/1.1.8)	Patient feedback from peer review and in the clinic	See above
003	SCM1	Ongoing pain management	Causes distress if suboptimal. Many wards do not have resources to meet needs for ongoing analgesia in a timely manner. May prolong admission to hospital.	Patient controlled analgesia is not always available even in large centres.	Peer review feedback
004	SCM1	Patient observations and recognition of acute chest complications	Failure to record observations may lead to delays in recognising over sedation with opiate drugs and development of acute chest syndrome. May be fatal if left untreated,	Data from audits and personal experience show that hypoxia is often not recognised or managed appropriately.	NCEPOD report
005	SCM1	Training of staff professionals in acute sickle cell crisis	Negative attitudes are a lack of experience impact on the quality and safety of patient care for this group of patients who may experience frequent admissions.	Patient feedback highlighted this as a major concern. Patients do not feel safe and often encounter negative attitudes from staff. This may in turn lead to fear of going to hospital or behaviours from the patients which reinforce negative perceptions by staff.	Feedback from patients
006	SCM2	Key area for quality improvement 1 All patients presenting to hospital with an acute painful sickle cell should be	Acute painful sickle crises are the cardinal feature of sickle cell disease. Despite this they are poorly managed and patient feedback continually highlights poor and inequitable management. Lack of adequate, rapid	The peer review programme of services for adults with haemoglobin disorders (2013-4) found large discrepancies in time to analgesia between different units. Most (but not all) units had recently audited this, but very few achieved this target. Once of most	The Overview report for the peer review process is being prepared and should be published within the next month. The Quality Standards for the

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
		offered analgesia within 30 minutes and pain control should be assessed every 30 minutes until satisfactory pain relief has been achieved and at least every 4 hours thereafter and	pain relief has a huge impact on patient experience and disempowers patients. It leads to a breakdown in the patient's relationship with the care providers and in the long term can lead to conflict which can further detriment patient care. Patients often avoid coming to hospital because of previous experience of long waiting times for analgesia and this can be damaging to their health. Even when initial analgesia is given within 30 minutes it is essential that patients have ongoing assessment to assess adequacy of analgesia and to ensure repeated doses of analgesia once pain relief wears off and pain returns. This is really a basic tenet of caring for any patient with pain, but patient feedback gives evidence that this is not done adequately.	consistent features of patient feedback on this programme was the long waits for analgesia experienced by patients in A+E departments. The peer review programme also found large discrepancies rates in frequency of re-assessment of pain between different units. This was not audited as frequently as 'time to analgesia, but where it had been audited few units achieved this target. Lack of reassessment of pain was a consistent features of patient feedback on the peer review programme	programme and all the individual reports are available on the West Midlands Quality Review Service website. http://www.wmqrs.nhs.uk/review-programmes/view/adults-with-haemoglobin-disorders
007	SCM2	Key area for quality improvement 2 Use of patient-controlled analgesia should be considered in all patients presenting to hospital with acute painful sickle crisis if repeated bolus doses of strong analgesia are needed within 2 hours. There should be locally agreed protocols for their use.	PCA is an effective way of providing pain relief for patients with acute sickle crisis and should be available for all patients if they wish to use it. There is clear inequity of care with some units being able to offer this service, but many other not being able to. PCA not only enables the patient to take control of their analgesia and empowers them, it is also cost effective.	.Despite good evidence that PCAs are effective they are still not available for patients with sickle cell disease in many hospitals around the country. This leads in many cases to inadequate pain relief being provided for patients and is detrimental for patient care. Even when PCAs are available they are often not available at all times of the day/week or in all of the hospital wards.	Am J Hematol. 2007 Nov;82(11):955-60. Patient-controlled analgesia versus continuous infusion of morphine during vaso-occlusive crisis in sickle cell disease, a randomized controlled trial. van Beers EJ, van Tuijn CF, Nieuwkerk PT, Friederich PW, Vranken JH, Biemond BJ.

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
008	SCM2	Key area for quality improvement 3 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 and attitudes towards and preconceptions about patients presenting with an acute painful sickle cell episode	Adequately trained staff are essential to ensure adequate pain relief is given promptly and monitored properly. Sickle cell patients can also develop complications secondary to or independently of acute painful crisis. These include acute chest syndrome, acute stroke, aplastic crisis. These have a high morbidity and mortality. It is essential therefore that staff are aware that they occur commonly in patients with sickle cell disease, monitor patients for these complications and provide early treatment.	The NCEPOD report into sickle cell disease gave several examples where inadequacy of monitoring following high doses of opioid analgesia contributed to patient death. The NCEPOD report (and other studies) have shown that the majority of deaths in patients with sickle cell disease are due to acute stroke and acute chest syndrome. Morbidity and mortality from these conditions can be reduced by early appropriate intervention. The peer review programme of services for adults with haemoglobin disorders found that training of health care professionals was inconsistent, and was not offered regularly or repeatedly in the majority of units reviewed even when they were caring for large numbers of patient with sickle cell disease. Patient feedback frequently said that staff did not have a good knowledge of sickle cell disease and its complications.	NCEPOD report. 'A sickle crisis?' The Overview report for the peer review process is being prepared and should be published within the next month. The Quality Standards for the programme and all the individual reports are available on the West Midlands Quality Review Service website. http://www.wmqrs.nhs.uk/review-programmes/view/adults-with-haemoglobin-disorders
009	SCM2	Key area for quality improvement 4 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 crisis	Day care settings are an effective way of providing initial assessment and treatment for acute painful sickle crises. Because staff in these settings have specialist knowledge patients receive rapid and appropriate analgesia and are assessed for sickle complications. Provision of early, effective treatment certainly leads to improved patient experience and improved pain control	The peer review programme of services for adults with haemoglobin disorders found that day unit facilities are not available in many units in England, even those with large sickle cell populations. Even where day units are available, opening hours are often restricted. Day unit services were highly rated by patients and had high levels of patient satisfaction.	Day case management of sickle pain: 3 years experience in a UK sickle cell unit. Wright J, Bareford D, Wright C, Augustine G, Olley K, Musamadi L, Dhanda C, Knight C. Br J Haematol. 2004 Sep;126(6):878-80. The Overview report for the peer

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
		should be available	and also results in decreased hospital admissions and length of stay		review process is being prepared and should be published within the next month. The Quality Standards for the programme and all the individual reports are available on the West Midlands Quality Review Service website.
010	SCM2	Key area for quality improvement 5 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.	Patients with sickle cell disease can present to any emergency department in England. In low prevalence areas particularly this may not be a very frequent occurrence and staff may not have much experience of management of sickle cell patients. It is especially important that these units have access to local protocols so there are clear guidelines for immediate care and that they have support from specialist centres for the care of complicated cases.	The peer review programme of services for adults with haemoglobin disorders found that even emergency departments in high prevalence areas did not always have locally agreed protocols for the care of acute painful crisis and sometimes although protocols may have existed, staff were not aware of them. This lead to delays in patients receiving adequate analgesia. The majority of centres in low prevalence areas were not reviewed but patient feedback would suggest that many hospitals do not have local treatment protocols and that this leads to significant delays in care. Emergency departments in low prevalence areas were not always aware of where to get specialist support from, especially out of hours. Furthermore specialist centres were not sure which hospitals they should be covering and did not provide a 24/7 service for advice.	The Overview report for the peer review process is being prepared and should be published within the next month. The Quality Standards for the programme and all the individual reports are available on the West Midlands Quality Review Service website.
011	SCM3	Key area for quality	Provision of appropriate and timely	The 2012 guideline is predicated on the	A comparison of the clinical

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
		improvement 1	analgesia in children. 1. Evaluation tools for pain. 2. Analgesic choices in children. 3. Education (for staff and carers) around pain evaluation and control within the setting of an acute sickle crisis.	provision of the best and most appropriate management of acute severe pain during a crisis. Quality improvements in this area are likely to have important impacts on embedding training and education of staff on the evaluation and management of acute pain in children with sickle cell disease. Both within and beyond the scope and settings of the guideline (home, schools, GP etc). 1. There is a large body of evidence available regarding the evaluation of pain in children, suggesting likely significant differences across disciplines and departments/units about which tools are most appropriate for evaluating childhood pain and who should evaluate pain (eg: self-reporting or by observation). This is particularly problematic in the very young or cognitively impaired eg: use of pain tools but also use of sedation score tools. 2. In practice analgesic choices in children tend to be limited to a small number of drugs within the major classes. This is despite a wealth of adult data on analgesic choices, a large body of work on pain in childhood from other disciplines (such as oncology or post-operative care) and the need to ensure that analgesic choices are tailored to the individual in terms of analgesic effect and adverse event profile. Absence of clarity on this area leaves children exposed to reduced choice, lack of RCTs comparing	utility of pain assessment tools for children with cognitive impairment. Voepel-Lewis T, Malviya S, Tait AR, Merkel S, Foster R, Krane EJ, Davis. Anesth Analg. 2008;106(1):72. Discordance between self-report and behavioral pain measures in children aged 3-7 years after surgery. Beyer JE, McGrath PJ, Berde CB. J Pain Symptom Manage. 1990;5(6):350.

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
				different agents and difficulty delivering on the guidelines section 1.11-1.17 regarding personalised care plans and failure to address specific areas such as managing adverse events without also reversing analgesic control. Specific drug limitations in young children (eg: use of codeine restricted in age<12y and failure of effect of codeine in some individuals) and variations in practice regarding access to PCAs and NCAs for the paediatric population are just some examples to barriers in place. 3. Education regarding managing pain during a sickle crisis; accessible to professionals and carers alike and taking into account the special needs of the very young or the cognitively impaired would be instrumental in developing uniformly high standards regardless of geographical location and reinforce measure around personalised care plans in which the patient or carer is truly regarded as an expert in their condition.	
012	SCM3	Key area for quality improvement 2	Delivering care (analgesia) in a timely way. This is an area for quality evaluation because of the impact on both clinical outcomes and patient experience.	Much of the guideline is around the delivery of care within a specific time frame alongside regular patient re-evaluation. This invites further exploration around: 1. Initial triage; should patients in sickle crisis be routinely triaged as 'urgent'? Do all units likely to be used by patients in crisis have a mechanism for triage? Is the same triage system in use across the	Manchester triage system in paediatric emergency care: prospective observational study. van Veen M, Steyerberg E, Ruige M *et al* (2008) *British Medical Journal*. Sep 22, 337

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
				UK, such as the Manchester Triage System? To what degree do units vary across the UK – such as direct access to day hospitals versus use of A&E services? Are services organised in such a way to ensure guidelines around timing of initial and subsequent evaluations to be realistically delivered? Are special measures in place for the very young or those who are cognitively impaired? 2. Specific work on ensuring open communication with patients and their carers around efficacy of pain control, reasons for not administering analgesia within the recommended time frames. There is often little work done to establish why clinical teams depart from guidelines and whether that departure is due to clinical reasons or operational reasons (eg: some units require anaesthetists to set up PCAs, or some patients decline analgesia within the designated time frame) 3. Patient feedback exercises. Both to ensure patients are being listened to in a manner consistent with the recommendations and to encourage patient involvement in the development and delivery of care	Reliability and validity of triage systems in paediatric emergency care. van Veen M, Moll HA. Scand J Trauma Resusc Emerg Med. 2009 Aug 27;17:38 Sickle crisis audit tool associated with the NICE guideline – exploring reasons why there may departure from the audit standards
013	SCM3	Key area for quality improvement 3	Personalised care and patient held records	The use of personalised care plans and the empowerment of patients and carers to be viewed as experts in their care is central to the guidelines. It opens the way to truly addressing patient concerns, strengthening	Little good quality published data?? Note: DoH Information Strategy 2012

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
				patient-clinician trust and ensures patient needs and values are central to coordinated and intelligent care delivery. Improvement in this area can only truly occur if information governance around how to access and share such information regardless of patient location (especially if held electronically) is examined in greater depth. This is particularly important regarding maintaining care plan accuracy (especially relevant for those patients who move frequently or do not engage regularly with outpatient services) and sharing sensitive information that may be relevant eg: safeguarding issues	
014	SCM3	Key area for quality improvement 4	Adverse events and safety	Much of this is covered in the above points on analgesic provision, choice, timing, evaluation and education around managing a patient in sickle crisis. Even so, specific attention should be paid to adverse event evaluation within a sickle crisis (sharing information in order to learn? SAE reporting?). Concerns over adverse events when using powerful analgesics in children are a contributory barrier to the provision of prompt strong analgesia or to considering alternatives when 1st line medications are not effective. In the very young and the cognitively impaired, evaluating pain (in groups who are not able to communicate pain intensity) exposes these groups to the risk of missed diagnosis of a different diagnosis or escalating pain suggesting a deterioration in crisis severity and/or the presence of a life threatening complication.	A Sickle Crisis? A report of the National Confidential Enquiry into Patient Outcome and Death (2008)

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
015	SCM4	Key area for quality improvement 1 Early Death Improvement	The Nice Guidelines show that people with Sickle cell are prone to Early death. More research and training of health workers will improve health conditions.	Many people suffer early deaths unnecessarily mainly due to insufficient knowledge on sickle cell.	There have been many incidences within the past few years showing carelessness of health workers who have not received training or knowledge on what sickle cell is. http://publications.nice.org.uk/treating-acute-painful-sickle-cell-episodes-in-hospital-ifp143/possible-complications
016	SCM4	Key area for quality improvement 2 Disability Status	Disability caused by strokes, organ failure and pneumonia can be conditions caused by sickle cell. A person's quality of life needs to be considered by health and social workers.	Presenting at hospital with a crisis can be due to the person experiencing housing issues, yet the social, psychological and wellbeing of the person are rarely considered before discharge. Government health and social care document states	Nice guidelines show that these issues may lead to readmission. Also NCPOD report http://www.ncepod.org.uk/2008report1/Downloads/Sickle_report.pdf http://www.legislation.gov.uk/ukpga/2012/7/enacted
017	SCM4	Key area for quality improvement 3 Race Equality	As Sickle Cell is mainly found in Black, Afro-Caribbean communities. It appears there is little to no evidence on government awareness campaigns in the U.K	People can only make an informed choice if they are given the information. The Sickle Cell population have been called "hard to reach" yet more visibility campaigns will help patients and public alike.	Please see http://sct.screening.nhs.uk/ More visible campaigns needed. http://www.legislation.gov.uk/ukpga/2012/7/enacted
018	SCM4	Key area for quality improvement 4 Hospital Stay VS Specialised Centre	Sickle Cell has been known about in the West for over 100 years, yet there are still insufficient places of specialised care in high prevalent areas. Nice Costing Report on Sickle Cell crisis has little evidence to show that specialised centres will cost less than hospital admissions. More investigation	People with a greater knowledge on how sickle cell crisis can damage the body need to train others in order for specialised areas of care to be established in high Sickle populations	http://www.nice.org.uk/nicemedia/live/13772/59767/59767.pdf

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
			needed.		
019	SCM4	Key area for quality improvement 5 Prioritise Nice Sickle Cell Guidelines in hospitals and clinics	Having several admissions to hospital recently, and accompanying others for admission. The 30 minute Nice guideline recommendation for pain relief had not been met on occasions.	Prolonged pain can cause bone and tissue damage as well as unnecessary discomfort to the individual experiencing the painful sickle cell crisis. More highlighting to health workers and hospital staff on Nice the existence of the Sickle Cell Guidelines.	NICE clinical guideline 143
020	SCM5	Key area for quality improvement 1 **All patients should be offered appropriate analgesia within 30 minutes of presentation**	Sickle cell pain can be variable in both severity and duration and appropriate analgesia is seen as the mainstay of sickle cell crisis care. Based on NICE and national guidance analgesia should be offered within 30 minutes of presentation	There is great variation throughout the UK in the management of pain for individuals with SCD with some patients having to wait for long periods of time for adequate analgesia (NCEPOD).	Rees DC, Olujohungbe AD, Parker NE, et al; British Committee for Standards in Haematology. Guidelines for the management of the acute painful crisis in sickle cell disease. Br J Haematol. 2003;120:744-52. Sickle Cell Disease in Childhood: Standards and guidelines for clinical care 2nd edition October 2010 A sickle cell crisis?, NCEPOD, 2008
021	SCM5	Key area for quality improvement 2 **Assessment and reassessment of pain using a validated, age appropriate pain tool**	Analgesia administration guidelines are based upon pain scores. To ensure adequate analgesia upon presentation and for ongoing management pain scores need to be undertaken using a validated, age appropriate pain tool. Reassessment of pain is recommended by NICE to be undertaken every 30	Initial assessment of pain is often undertaken, although there is not always documentation of a validated tool being used. However ongoing pain assessment is often inadequate and not performed often enough or according to medical instruction.	Rees DC, Olujohungbe AD, Parker NE, et al; British Committee for Standards in Haematology. Guidelines for the management of the acute painful crisis in sickle cell disease. Br J Haematol. 2003;120:744-52.

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
			minutes until satisfactory pain relief has been achieved and then at least every 4 hours thereafter.		Royal College of Paediatrics and Child Health. Guidelines for good practice: recognition and assessment of acute pain in childhood. London: RCPCH; 2001. A sickle cell crisis?, NCEPOD, 2008
022	SCM5	Key area for quality improvement 3 **Individualised care pathways for individuals presenting with a sickle cell crisis**	Each sickle cell patient is unique and become experts in their own/their child's pain.	NCEPOD recommend that healthcare professionals work with patients to develop individualised pain management strategies	A sickle cell crisis?, NCEPOD, 2008
023	SCM5	Key area for quality improvement 4 **All healthcare professionals who care for patients with an acute painful sickle cell episode should receive regular training**	A major complaint of patients is that health care professionals managing their care do not have sufficient knowledge about sickle cell disease.	Managing a painful crisis can be challenging. Health care professionals need expert knowledge in order to provide more effective and comprehensive care for patients with SCD To provide equity of care. Patients should be able to receive the same standard of care which ever hospital they attend.	Sickle Cell Disease in Childhood: Standards and guidelines for clinical care 2nd edition October 2010 Caring for people with sickle cell disease and thalassaemia syndromes: A framework for nursing staff RCN 2012 Understanding the contribution of sickle cell and thalassaemia specialist nurses. 2012 A sickle cell crisis?, NCEPOD, 2008

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
024	SCM5	Key area for quality improvement 5 **Patients/carers are offered verbal and written information on how to manage their current crisis episode as part of the discharge process.**	To fully inform individuals/carers on how to continue to effectively manage their pain whilst at home on informing them of any symptoms that they must reattend hospital for. Individuals can be discharged home whist still in crisis. Individuals often develop worsening of a crisis at day 5. If this is at home then patients may need readmission to hospital.	To allow individuals to manage their sickle cell in partnership with health care professionals. Education and support for individuals and carer's is an essential part of ongoing sickle cell care	Sickle Cell Disease in Childhood: Standards and guidelines for clinical care 2nd edition October 2010
025	SCM6	Key area for quality improvement 1	There is evidence that managing acute pain in sickle disease well Is essential	Reducing the delay in receiving appropriate medication .	NICE has published guidelines to support the importance of pain management in the acute crisis
026	SCM6	Key area for quality improvement 2	Seamless integration of pain management between out and inpatient episode improves patient experience	Managing the condition in the longterm as survival rates are increasing, with a view of reducing reliance on opioid medication	NICE patient experience guidelines
027	SCM6	Key area for quality improvement 3	Research : Effectiveness of non opioid medication for pain	Reducing opioid medication reduces longterm side efeffects such as hyperalgesia, immunesuppression and hormonal changes	British Pain Society guidelines on the use of opioid medication
028	SCM6	Key area for quality improvement 4	Trial network and registry	Advancing new treatment such as haematopoetic transplantation	In cancer care almost all patients are entered into multicentre trials (national/international trialnetworks) and progress in treating various cancers is advancing fast
029	UK Forum on Haemoglobin Disorders	Key area for quality improvement 1 **Appropriate pain**	**Specifically: Pain assessed promptly after presentation, and timely, appropriate and effective analgesia administered.**	Pain in acute sickle crisis can be of excruciating severity, and is often underestimated and poorly managed in hospital, causing undue suffering to the	NCEPOD report 2008 ‘A sickle cell crisis?’

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
		management	There is evidence for variable management of acute pain, with some underprescribing / delays and some overprescribing with adverse outcomes.	patient, and sometimes adverse effects from over-enthusiastic strong analgesia.	
030	UK Forum on Haemoglobin Disorders	Key area for quality improvement 2 **Complete and sufficiently frequent recordings of physiological observations throughout, with escalation of abnormal findings,**	**Specifically: Complete sets of physiological observations recorded at presentation, and intervals after according to the measures: suggest according to National Early Warning Score observation chart, with appropriate escalation to higher levels of care [Progressive Care / Intensive Care] if significant abnormalities.** Patients presenting with apparently uncomplicated crisis can develop a range of additional problems / sepsis / organ failure and accurate, sufficiently frequent physiological observations from the basis for identifying deterioration and escalating to appropriate clinical staff to allow necessary additional interventions.	There are many cases – reported through adverse event recording on National Haemoglobinopathy Registry, presented at UK Forum for Haemoglobin Disorders scientific meetings, and from hospital Mortality and Morbidity reviews – of 'failure to rescue' or to adequately recognise how seriously ill patients are / become while in hospital, leading to adverse outcomes. This is in acute medical patients in general, and relevantly in those with sickle cell crisis / additional complications also.	NCEPOD report 2008 'A sickle cell crisis?' National Institute for Health and Clinical Excellence. *Acutely ill patients in hospital. Recognition of and response to acute illness in adults in hospital*. NICE clinical guideline 50. London: NICE, 2007. National Confidential Enquiry into Patient Outcome and Death. *Emergency admissions: a journey in the right direction?* London: NCEPOD, 2007. Royal College of Physicians. *Acute medical care: the right person, in the right setting – first time*. London: RCP, 2007. Royal College of Physicians. *National Early Warning Score [NEWS]; standardising the assessment of acute-illness severity in the NHS.* 2012
031	UK Forum on Haemoglobin Disorders	Key area for quality improvement 3 **Vigilance for the presence or development of additional complications**	**Specifically: Frequent clinical assessment throughout the admission by physical examination, complete physiological observation recordings, referring to results of**	Morbidity and mortality result from missed infection, renal or hepatic impairment, developing chest syndrome. These may be manifest only after presentation with apparently uncomplicated pain crisis so	Boundless published and unpublished case reports. NCEPOD report 2008 'A sickle cell crisis?'

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
		underlying pain crisis.	**haematological, biochemical, radiological investigations to allow early identification of additional complication on top of pain, such as acute severe anaemia, infection, specific organ involvement.** It cannot be assumed that a presentation with acute pain crisis is uncomplicated, and a variety of serious additional complications may underlie the presentation or may develop during admission. Careful vigilance is required to ensure these are identified and managed promptly.	careful ongoing surveillance is essential.	
032	UK Forum on Haemoglobin Disorders	Key area for quality improvement 4 **Use of blood transfusions in sickle cell crisis**	**Specifically: guidance on the use of red cell transfusions – additive or exchange transfusions – for different presentations and complications, requiring audit of use against these and ensuring adequate steps are taken to identify / avoid antibody formation.** Red cell transfusions can be life-saving in sickle cell crisis, for example in aplastic crisis with profound anaemia, chest syndrome, stroke. However, they are *usually* not indicated in uncomplicated pain crisis, and always carry a degree of risk to the recipient so should be avoided if not necessary.	Administration of ill-matched red cell units can lead to serious haemolytic transfusion reactions and occasionally death [for example, those which contain antigens to which the recipient has previously made an antibody reaction, even tho that may no longer be detectable in current compatibility tests]. Also: there is evidence of variable transfusion practice, acutely and longer term / planned transfusion, and anecdotal reports of patients receiving unnecessary transfusion eg for relatively minor falls in Hb, especially in hospitals less familiar with sickle cell and its complications. The risks and benefits of transfusion need to be carefully weighed in every individual case.	Annual 'Serious Hazards of Transfusion' reports ['SHOT'], many case reports, and findings of Peer Review visits to services for both children and adults with these conditions [available through West Midlands Quality Review Service website].
033	UK Forum on Haemoglobin	Key area for quality improvement 5	Most pain episodes are managed by patients and their families or carers at	Patients – especially those with frequent pain crises – regularly express the wish to be	Patient groups feedback on services.

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	Disorders	**Support for management of uncomplicated pain crisis at home.**	home, and they self-evaluate when they can no longer manage and need hospital care. There is evidence that even more severe pain crises, which have no additional or underlying complications, after adequate assessment and if appropriate analgesia is available for home use.	managed at home where possible, where they feel they recover more quickly and without the disturbance and exposure to other risks that hospital care engenders. Community services / outreach hospital services can improve patient experience in this way, while also releasing acute hospital beds for times when patients have additional complications requiring acute medical input.	Prolonged experience of outreach services in NE London. Experience of Community care in Camden. Current research project at Barts Healthcare formally evaluating home care.
034	Napp Pharmaceuticals Limited	Key area for quality improvement 1 All sickle cell patients to carry up to date patient information cards detailing current management including type of analgesia and dose	Patients need to have their acute episodes of pain controlled fast and efficiently when presenting at hospital. Existing medication needs to be taken into account as this background level of analgesia could be higher than locally recommended starting doses for pain in general and may lead to unnecessary delays in reaching optimal pain control. The cards may also help to identify legitimate patients.	Patients will be in acute severe pain and any delay in treating them with an adequate and appropriate dose will lead to unnecessary pain, anxiety and potentially to complications. Patients may be away from their own home hospital and therefore may not be known to the staff at the hospital unit to which they have gone. No national sickle cell patient data base or treatment protocol exist..	
035	Napp Pharmaceuticals Limited	Key area for quality improvement 2 Each and every A&E and medical department to have and to demonstrate up to date protocols for the management of sickle cell crises including pain management	It is important that all healthcare professionals within these departments give consistent patient care and do not overlook key aspects of sickle cell crisis when planning individual patient care. It is also important that patients receive the most appropriate medicine to control pain in A&E, continuing good pain control if admitted to a ward and on discharge.	Nationally there is no agreed protocol for the treatment of sickle cell crisis. NICE has produced a guideline (CG143) but this needs to be implemented at local level. The NICE guideline should form the basis for the development of a treatment protocol incorporating local formulary and choice of medicine. Any pathway / protocol should encompass A&E / admittance to hospital ward and discharge.	Sickle cell acute painful episode (CG143)

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036	Napp Pharmaceuticals Limited	Key area for quality improvement 3 All medical and A&E departments should be able to demonstrate that all staff have been adequately trained in the assessment and treatment of sickle cell crises including the assessment of pain.	Assessment is key to good pain management. Furthermore with junior hospital staff often working on shifts and on rotation it is imperative that they receive training to ensure consistent and specific training on the management of pain associated with sickle cell crises. The average length of undergraduate training on pain received by doctors is on average only 13 hours. (Briggs et al 2011)	The level of training in pain management has been the subject of a number of publications. The amount of time veterinary surgeons receive is twice as long as that received by doctors. Furthermore pain is one of the main reasons that patients present to A&E. It is important that doctors can identify, assess and manage the complications of acute pain, chronic pain with acute flare ups (commonly referred to as breakthrough pain). Drs deal with many patients who have a poor understanding of English or who may communication problems. The use of the most appropriate pain assessment tools should be encouraged within the QS and if necessary these could be provided in languages other than English.	http://www.ncbi.nlm.nih.gov/pubmed/21330174 Briggs et al Eur J Pain 2011 15 (8) 789-95 http://www.britishpainsociety.org/pub_pain_scales.htm
037	Napp Pharmaceuticals Limited	Key area for quality improvement 4 That all patients with sickle cell crises are administered the most appropriate analgesia in the shortest possible time frame	Patients presenting with sickle cell crisis need to have fast access to pain medication which will have a quick effect on their pain. Oral medication offers a fast and easy solution and allows staff the opportunity to assess progress before initiating costly PCA medications which requires the availability of equipment and staff trained in its use. It also has the disadvantage of the patient being more dependent on nursing staff. Patients will also need to be informed of how to use the PCA and although not	It is important for patients to be managed efficiently with the most appropriate medication. The British Pain Society has a booklet of recommendations on prescribing opioids for chronic pain. The information here is also useful background for prescribing opioids in general.	http://www.britishpainsociety.org/book_opioid_main.pdf

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			onerous it may be difficult in times of stress and anxiety for the patient to understand.		
038	British Pain Society	Key area for quality improvement 1 **Timely and expert assessment of acute pain during sickle cell crisis (SCC)**	NICE 2012 guideline on acute sickle cell crisis recommends: *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.* *1.1.6 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.* *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.*	Patients with known sickle cell disease (SCD) should be seen within 30mins in A&E or on a haematology unit by a specialist in haematology/SCD or a pain specialist trained in SCD. If acute pain is not recognised promptly it may lead to unnecessary suffering; inappropriate discharge from A&E for the patient only to return again; inappropriate initiation of inadequate medication; inappropriate initiation of invasive medication when it is not needed. Inappropriate or inexpert use of opioids in A&E or acute medical admitting units carries high risk of opioid toxicity including respiratory depression. Some patients may present with non-SCD pain that is mis-diagnosed as SCC.	Sickle cell acute painful episode (CG143)
039	British Pain Society	Key area for quality improvement 2 **Timely initiation of multimodal pharmacological management of acute**	NICE 2012 guideline on acute sickle cell crisis recommends: *1.1.4 Offer analgesia within 30 minutes of presentation to all patients presenting at* *hospital with an acute painful sickle cell*	If SCD pain is not treated promptly and expertly using multimodal analgesia (opioid, NSAID ± benzodiazepines for anxiety and ± gabapentin or pregabalin for co-existing neuropathic pain) then there is a risk of pain escalating and necessitating admission for	Sickle cell acute painful episode (CG143)

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		pain in SCC	*episode (see also recommendations*	more invasive management.	
040	British Pain Society	Key area for quality improvement 3 **Specialist follow-up of patients discharged after hospital admission requiring strong opioid treatment**	NICE 2012 guideline on acute sickle cell crisis recommends: *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.* *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.*	If patients have received strong opioids during hospital admission, and especially if they are discharged with opioid medication, then there is a risk of dependency behaviour and possibly addiction, if the patient is not followed up by a specialist experienced in managing SCD.	Sickle cell acute painful episode (CG143) BPS documents on opioids for chronic pain
041	British Pain Society	Key area for quality improvement 4 **Use of non-pharmacological interventions and psychological self-management during and in recovery from acute SCC**	NICE 2012 guideline on acute sickle cell crisis recommends: *Non-pharmacological interventions* *1.1.22 Encourage the patient to use their own coping mechanisms (for example, relaxation techniques) for dealing with acute pain.*	Over-reliance on drug management may be detrimental to some patients, especially those with underlying psychological disorders such as anxiety and depression. If patients are assessed for psychological needs and referred appropriately this could lead to reduced drug dependency and addiction.	BPS documents on chronic pain
042	The Royal College of Pathologists and	Key area for quality improvement 1	Analgesia	Effective, safe and acceptable opioid protocol	NICE clinical guideline, Cochrane review of pain in sickle cell disease, NCEPOD

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	British Committee for Standards in Haematology				report, UK standards of care for children and adults with SCD, NHS recommendations on dosing and avoiding overdose of opioids in naïve patients
043	The Royal College of Pathologists and British Committee for Standards in Haematology	Key area for quality improvement 2	Safety	Regular observations at least of pain score, respiratory rate and sedation score on an hourly basis during first six hours of attendance in emergency department with acute pain	NICE clinical guideline, Cochrane review of pain in sickle cell disease, NCEPOD report, UK standards of care for children and adults with SCD, NHS recommendations on dosing and avoiding overdose of opioids in naïve patients
044	The Royal College of Pathologists and British Committee for Standards in Haematology	Key area for quality improvement 3	Environment	Designated clinical area/s which are conducive for recovery from acute pain crisis	NICE clinical guideline, Cochrane review of pain in sickle cell disease, NCEPOD report, UK standards of care for children and adults with SCD
045	The Royal College of Pathologists and British Committee for Standards in Haematology	Key area for quality improvement 4	Staff education	Awareness of front line and specialised medical and nursing staff on clinical features of sickle cell crises, how to manage acute pain with local protocol and how to understand and communicate with patients in a sensitive and caring fashion	NICE clinical guideline, Cochrane review of pain in sickle cell disease, NCEPOD report, UK standards of care for children and adults with SCD
046	The Royal College of Pathologists and British Committee for Standards in	Key area for quality improvement 5	Management of acute on chronic pain and of opioid overuse in frequent hospital attenders	Identification of alternative ways of managing long-lasting pain (as an alternative to injected opioids), better liaison with community support services, chronic pain, specialist psychology and voluntary organisations to support home management with non-injected	NICE clinical guideline, Cochrane review of pain in sickle cell disease, NCEPOD report, UK standards of care for children and adults with SCD

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	Haematology			analgesics, and non-pharmacological approaches to pain management	
047	Nordic Pharma Ltd	Key area for quality improvement 1 Ensure patients who have been discharged following a crisis have a follow up appointment made as outpatient or with their Sickle Cell specialist centre or have an appointment made with a specialist centre if this is a new patient.	There are a number of specialist centres for Sickle Cell disease around the UK, the hospital treating the patient for the crisis might not be one of these centres. Thought should be given to providing a quality standard to ensure the appropriate follow up and ongoing management of these patients in upon discharge from hospital for example	Appropriate ongoing treatment, counselling by staff at specialist Sickle Cell centres or by a Specialist Haemoglobinopathy Team or consultant might reduce the incidence of further episodes and admissions to hospital.	BCSH Guidelines for the Management of Acute Painful Crisis in Sickle Cell Disease. Br J Haematol 2003, 120. 744-752. http://www.bcshguidelines.com/documents/sicklecelldisease_bjh_2003.pdf Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK 2008. Sickle Cell Society. http://www.sicklecellsociety.org/app/webroot/files/files/CareBook.pdf Standards for adult sickle cell disease aim to reduce gaps in care. Mayor S. BMJ 2008; 337:132 http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2483891/
048	Nordic Pharma Ltd	Key area for quality improvement 2 Discharge letters should be sent to the GP within 10 working days of hospital discharge following an acute painful crisis	This is to ensure continuity of care between secondary and primary care once a patient is discharged	The current guidelines as such don't recommend this. To ensure this continuity of care takes place we would suggest a quality standard be set to reflect the Sickle Cell Society guidelines - that discharge letters be sent to the patient's GP.	Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK 2008. Sickle Cell Society. http://www.sicklecellsociety.org/app/webroot/files/files/CareBook.pdf

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					Standards for adult sickle cell disease aim to reduce gaps in care. Mayor S. BMJ 2008; 337:132 http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2483891/
049	Nordic Pharma Ltd	Key area for quality improvement 3 There appear to be no standards for the prevention of sickle cell crises or the management of chronic long term management. A section on prevention of further crises or a separate standard/guideline could be something to consider.	There is currently no place in these standards for the prevention of further crises or the ongoing chronic management of these patients. It is recommended in the UK's Sickle Cell Society Guidelines that there be improved prevention in primary care The NICE guideline also recognise that the prevention of some complications form SCD is important from an economic as well as patient care perspective	Whilst these standards focus on care of a crisis there should be thought given to the prevention or minimisation of risk of further crises or complications. It has been suggested that the chronic nature of sickle cell disease has been insufficiently recognised. The NICE guideline also recognise that the prevention of some complications form SCD is important from an economic as well as patient care perspective	Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK 2008. Sickle Cell Society http://www.sicklecellsociety.org/app/webroot/files/files/CareBook.pdf Standards for adult sickle cell disease aim to reduce gaps in care. Mayor S. BMJ 2008; 337:132 http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2483891/ Sickle cell acute painful episode: Management of an acute painful sickle cell episode in hospital. NICE: June 2012. http://www.nice.org.uk/CG143 Experiences of Hospital Care and Treatment Seeking for Pain From Sickle Cell Disease: Qualitative Study. Maxwell & Streetley BMJ 1999; 318:1585. http://www.bmj.com/content/318/

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					7198/1585
050	Sickle Cell Society	Management of acute painful sickle cell episodes	There is recent evidence(hospital peer reviews) that shows the management of acute painful sickle cell episodes for patients presenting at accident and emergency hospital departments and within hospital wards, continues to be variable throughout England. Specific issues include delays in receiving analgesia and inappropriate analgesia.	It is an important area for quality improvement because it is a consistent source of complaints and frustration from individuals with sickle cell and their families. It also raises important questions about the level and quality of care provided for this group of patients. Many patients and family members are expert in their condition. Since sickle cell is a long term condition and because of the episodic nature of crises, the variability in the quality of care is a serious cause of concern.	Emerging findings from recent first round of hospital peer reviews. Formal and anecdotal feedback and complaints to the Society from individuals with sickle cell and their families.
051	Sickle Cell Society	Staff knowledge and understanding of sickle cell	Variability of management of management of acute painful crises also relates to the knowledge and understanding of clinical staff. For example presentations to accident and emergency outside of hospital sickle cell centre clinic times have shown that some staff lack knowledge and understanding of the management of the condition which is exacerbated by lack of care management plans or guidance.	Care can be inconsistent with different health professionals. The specific issues include understanding, awareness and training of staff, staff recognising signs and symptoms and complications. This too is a source of complaints and frustration for individuals with sickle cell and their families. The Society believes strongly that patients and the Society can play an effective part in supporting training and development activities to promote better understanding and awareness of sickle cell in clinical and non clinical settings.	Formal and anecdotal feedback and complaints to the Society from individuals with sickle cell and their families. Annual patient education seminars
052	Sickle Cell Society	Managing pain in adolescent males	The current guideline is generic and as far as can be seen from a patient perspective makes no reference to priapism, although it does refer to management of acute painful sickle cell	Feedback suggests that for some adolescent males, this can be a difficult subject to discuss with health professionals. There is good evidence that priapism left untreated or indeed unrecognised can have	Westerdale 1997 Okpala 2002

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			episodes in children and young people.	other health implications such as impotency.	
053	Sickle Cell Society	Health education and Health promotion	There is good evidence to suggest that a holistic approach should be taken to sickle cell	The guidelines particularly in relation on-going management and discharge should signpost individuals to material (some of which can be provided by the Society) on pain analgesia, sexual health, psychosocial support, pregnancy, screening	
054	NHS England	Key area for quality improvement 1	Provision of pain relief in day care setting, ie outside accident and emergency departments. Ref 1.1.24 of Sickle pain guideline	NCEPOD 2008 mortality review highlighted failures in many aspects of acute care including urgent care; Increasing pressure on Emergency departments generally is likely to mean maintenance of an adequate standard of care will become more difficult. Evidence from patients shows a much higher satisfaction with services for Sickle pain when provided outside Accident departments by a team familiar with them and with Sickle	NCEPOD 2008 report into mortality Adult and Paediatric peer review data 2011 (paediatric) 2013 (adult) available on http://www.wmqrs.nhs.uk/, the latter suggests that initial provision of pain relief outside Emergency departments remains uncommon .
055	NHS England	Key area for quality improvement 2	Evidence of pain relief being provided as per NICE guidance within 30 minutes of presentation Ref 1.1.12 of Sickle pain guideline	This is a key recommendation in the NICE guideline on Sickle pain, slow and inadequate pain relief remains the single most common issue identified by patient groups.	Evidence in the form of audits of pain relief presented to both Paediatric and adult peer reviews suggests that this standard is not being met consistently. http://www.wmqrs.nhs.uk/
056	NHS England	Key area for quality improvement 3	Evidence that all Trusts have clear guidelines on the management of Sickle crisis as per NICE guidelines including recognition of more complex complications and clear escalation of such problems to specialist centres. REF 1.1.25 of Sickle pain guideline	NCEPOD 2008 report showed that non Sickle causes of pain or rarer complications of Sickle may not be recognised. The organisation of care into local and responsible specialist units via CRG specification means that all should have a clear access to protocols and a route to specialist expertise when needed, particularly	Evidence from NCEPOD 2008 , adverse event reporting via National Haemoglobinopathy Registry http://www.nhr.nhs.uk/

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				relevant to low prevalence areas.	
057	NHS England	Access to monitoring of % HbS during sickle cell crisis	In certain types of sickle cell crisis such as haemolytic crisis there is a need for exchange transfusion. In these cases monitoring of %HbS can provide important data that can guide treatment and improve outcomes	Not all haematology laboratories are able to provide a 24/7 or even 7/7 service for %HbS monitoring. This would be particularly important in areas with a significant prevalence of SCD. There is no clear standardisation of reporting between laboratories	There is no published evidence although this is deemed best practise from a number of scientific experts in high prevalence areas. (Level III evidence)
058	NHS England	Scientists as part of multi-professional teams caring for patients with SCD	To ensure appropriate laboratory provision, advice and interpretation of results	There needs to be a joined up approach to the laboratory support of sickle cell crisis to ensure results are available in a timely manner.	There is no published evidence although this is deemed best practise from a number of scientific experts in high prevalence areas. (Level III evidence)
059	National patient safety function of NHS England		We have not published evidence from the National Reporting and Learning System (NRLS) relating to sickle cell crisis, but reviewed data in support of the 2008 NCEPOD report this quality standard references.	Whilst we appreciate NRLS data cannot inform actual content of the standard, NICE may be able to use the following information in prioritising areas for inclusion: • NRLS reports confirm the importance of appropriate pain relief in sickle cell crisis, in terms of both avoiding inadequate pain relief and avoiding opiate overdosing • NRLS reports suggest criteria for treating young adult patients in adult or paediatric services need to be clear to avoid delays in treatment • NRLS reports suggest that in many hospitals haematology is a primarily outpatient service that may not have designated beds. Identifying a ward/unit	

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				where patients with Sickle cell crisis can be routinely admitted will help ensure staff have the right skills and awareness.	
060	Pfizer	No comments to make			
061	RCPCH	No comments to make			
062	RCN	No comments to make			