NATIONAL INSTITUTE FOR HEALTH AND CARE EXCELLENCE

HEALTH AND SOCIAL CARE DIRECTORATE

QUALITY STANDARD CONSULTATION

SUMMARY REPORT

1 Quality standard title

Sickle cell crisis.

Date of Quality Standards Advisory Committee post-consultation meeting:
16 January 2014.

2 Introduction

The draft quality standard for sickle cell crisis was made available on the NICE website for a 4-week public consultation period between 19 November and 17 December 2013. Registered stakeholders were notified by email and invited to submit consultation comments on the draft quality standard. General feedback on the quality standard and comments on individual quality statements were accepted.

Comments were received from 14 organisations, which included service providers, national organisations, professional bodies and others.

This report provides the Quality Standards Advisory Committee with a high-level summary of the consultation comments, prepared by the NICE quality standards team. It provides a basis for discussion by the Committee as part of the final meeting where the Committee will consider consultation comments. Where appropriate the quality standard will be refined with input from the Committee.

Consultation comments that may result in changes to the quality standard have been highlighted within this report. Comments suggesting changes that are outside of the process have not been included in this summary. The types of comments typically not included are those relating to source guidance recommendations and suggestions for non-accredited source guidance, requests to broaden statements out of scope, requests to include overarching outcomes, thresholds, targets, large volumes of supporting information, general comments on the role and purpose of quality standards and requests to change NICE templates. However, the Committee should read this summary alongside the full set of consultation comments, which are provided in appendix 1.

3 Questions for consultation

Stakeholders were invited to respond to the following general questions:

1. Does this draft quality standard accurately reflect the key areas for quality improvement?

2. If the systems and structures were available, do you think it would be possible to collect the data for the proposed quality measures?

4 General comments

The following is a summary of general (non-statement-specific) comments on the quality standard.

- Quality standard comprehensive, succinct and accurately reflects the key areas for quality improvement.
- The quality standard should extend to ambulance services and primary and community healthcare staff.

Consultation comments on data collection

- Data collection felt feasible for all statements.

5 Summary of consultation feedback by draft statement

5.1 Draft statement 1

People who present at hospital with an acute painful sickle cell episode have an assessment and appropriate analgesia within 30 minutes of presentation.

Consultation comments

Stakeholders made the following comments in relation to draft statement 1:

- Timely provision of analgesia should be highlighted as a multidisciplinary issue and include the role of pharmacists to provide advice and ensure clinical areas have access to medication.
- Stakeholders suggested the statement should outline the steps to identify quickly and effectively what pain relief each patient requires including the option of red cell exchange.

5.2 Draft statement 2

People with an acute painful sickle cell episode have an assessment of pain relief every 30 minutes until satisfactory pain relief has been achieved and then at least every 4 hours.

Consultation comments

Stakeholders made the following comments in relation to draft statement 2:

- Concern raised that reassessment every 30 minutes is difficult to achieve beyond the first 1-2 hours.
- ‘Satisfactory pain relief’ felt difficult to measure.
- Suggestion to align timescales with statement 3 after the first hour.

5.3 Draft statement 3

People with an acute painful sickle cell episode who are taking strong opioids are monitored for adverse events every hour for the first 6 hours after presentation and then at least every 4 hours.

Consultation comments

Stakeholders made the following comments in relation to draft statement 3:

- Suggestion the statement should be expanded to weaker opioids.
- Suggestion to remove blood pressure from the list of required assessments as repeated blood pressure measurements was highlighted to lead to sickling in the affected arm.
- Suggestion that monitoring should restart when analgesia is stepped up.
- Concern that the statements fall short of typical institutional guidelines for the management of children on strong opioids with no mention of paediatric acute pain teams.

5.4 Draft statement 4

People with an acute painful sickle cell episode are assessed for acute chest syndrome if they have 1 or more of the following: abnormal respiratory signs or symptoms, chest pain, fever, or signs and symptoms of hypoxia.

Consultation comments

Stakeholders made the following comments in relation to draft statement 4:

- Whenever SaO2 is measured the FiO2 (inspired oxygen) needs to be recorded.
- Outcome measure suggested on staff awareness about acute chest syndrome being a complication.
- Data collection issues raised due to the variety of documents and formats it may be recorded in. Suggested the use of a checklist.

5.5 Draft statement 5

People with an acute painful sickle cell episode are cared for by healthcare professionals who have access to locally agreed protocols for treatment and management and specialist support for sickle cell care from designated centres.

Consultation comments

Stakeholders made the following comments in relation to draft statement 5:

- Locally agreed protocols should be identified as a multidisciplinary issue with appropriate support from healthcare professionals including pharmacists and multidisciplinary pain clinics.

6 Suggestions for additional statements

The following is a summary of stakeholder suggestions for additional statements.

- Statement suggested on access to psychological therapies for pain management.
- Statements suggested on provision of discharge information and follow up by specialist or GP particularly for people discharged on strong opioids.
- Statement suggested on community care for people who have no support at home.
- Sickle cell crisis and pregnancy with reference to the RCOG guideline and awareness of not using NSAIDS in pregnancy after 28 weeks.
- Need for a care pathway from ambulance to post discharge.
- Statement for ambulance services so that ambulance staff follow set procedures for the management of sickle cell crisis.
- Need for evaluation of the standards to ensure they are being implemented.

Appendix 1: Quality standard consultation comments table

ID	Stakeholder	Statement No	Comments
1	Department of Health	General	Thank you for the opportunity to comment on the draft for the above quality standard. I wish to confirm that the Department of Health has no substantive comments to make, regarding this consultation
2	Napp Pharmaceuticals Limited	General	Thank you for the opportunity to comment on this draft of the NICE QS for Sickle Cell crisis
3	Napp Pharmaceuticals Limited	General	We have no further comments to add in relation to the details of each of the quality statements.
4	NHS England	General	Thank you for the opportunity to comment on the draft scope for the above quality standard I wish to confirm that NHS England has no substantive comments to make regarding this consultation
5	North West London Hospitals NHS Trust	General	NW London Hospitals NHS Trust Hospitals team have reviewed the document and apart from one or two typographical errors we have no specific comments. It is pretty comprehensive, succinct and all the standards are what we were familiar with already and are measureable.
6	Royal College of Nursing	General	This is to inform you that there are no comments to submit on behalf of the Royal college of Nursing to inform on the draft sickle cell quality standard consultation
7	Royal College of Paediatrics and Child Health	General	Thank you for inviting the Royal College of Paediatrics and Child Health to comment on the Sickle cell crisis draft standard. We have not received any responses for this consultation
8	Napp Pharmaceuticals Limited	Introduction	Napp are pleased to see that training and competencies are included and would encourage specific pain management related training.
9	Association of Paediatric Anaesthetists of Great Britain and Ireland	Question 1	Whilst we recognise that these quality standards are based on the recommendations within CG143 (June 2012), we are concerned that these recommendations fall short of typical institutional guidelines for the management of children on “strong” opioids via NCA, PCA or infusion. Routine nursing observations should take place hourly for the duration of analgesia: HR, BP, RR, and temperature, including hourly assessment of pain with an age appropriate tool, sedation and nausea and vomiting. This should be supplemented by continuous SaO2 monitoring, whether the patient is receiving supplementary oxygen or not. Supervision would be by a Paediatric Acute Pain Team, this is not mentioned in this Quality Standard, yet is a basic standard in many clinical standards documents. Some patients will require significant quantities of opioid, which may be beyond typical acute pain protocols. Some institutions will have bespoke protocols to cover this eventuality, with clear guidance with regard to threshold for HDU admission; this would be a useful Quality standard. Since the quality standard is superseded by many institutional guidelines, though compliance with previously

ID	Stakeholder	Statement No	Comments
			published guidance will be assessed, such compliance will not reflect an opportunity for quality improvement. Consideration should be given to a rewording of the quality standard to reflect common practice in UK institutions, (indicated above).
10	Central and North West London NHS Foundation Trust	Question 1	It is notable that there are no recommendations concerning psychological aspects of pain management in the review. In the Cochrane Review of 2012 Kofie and Green identified CBT as relevant for reducing the affective component of pain. (Cochrane Database Syst Rev. 2012 Feb 15;2:CD001916. doi: 10.1002/14651858.CD001916.pub2. Psychological therapies for sickle cell disease and pain.Anie KA, Green J.Source Brent Sickle Cell and Thalassaemia Centre, Imperial College Faculty of Medicine, London, UK. kofi.anie@nhs.net) Moreover the evidence for CBT in long term health conditions in general in considerable and should be considered relevant. Referral for longer term psychological input from the time of crisis should help with management of subsequent admissions. We would suggest that it constitutes an inequality to deny this group access to psychological therapy to help manage their crisis admission.
11	Faculty of Pain Medicine	Question 1	We propose to consider an additional quality standard on provision of discharge information (patient/GP) and specialist follow up, particularly for patients discharged on strong opioid analgesia. These patients should have a dose-reduction plan with timeline for complete withdrawal of strong opioids and a sole prescriber of opioids. Long-term use should be avoided as it can have major implications for the sickle cell sufferer, e.g. use of strong opioid analgesia between crises, development of opioid-induced hyperalgesia or endocrine changes etc.
12	Nordic Pharma Ltd	Question 1	Thank you for allowing us to comment on the draft Quality Standard. Nordic Pharma suggest this section could be expanded to include some standard on the quality of care once the patient is discharged. It is included in Sickle Cell guidelines that follow up appointments should be made and that discharge letters be sent to the patient's GP within 10 days following discharge from hospital. Although there is limited information available, there is some evidence to suggest that this is an area that could be improved on. According to a focus group study published in 2012, it is reported that patients feel it is important for their GP to be informed when they have attended hospital as a result of a Sickle Crisis in order to ensure adequate continuity of care. It is reported that this does not always happen. This study from North London outlining a patient focus group's view on improving sickle cell disease treatment found that there was seemingly a lack of cohesion and communication between the hospitals, GPs and specialist centres. The Focus group members emphasised that there was never a follow up from their GPs following hospital discharge. This could be incorporated into the standards to help ensure this continuity of care happens.

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			AlJuburi G, Phekoo KJ, Ogo Okoye NV et al, Patient's views in improving sickle cell disease management in primary care: focus group discussion. JRSM Short Report., 2012, v3(12): 1-7 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
13	Royal College of Obstetricians and Gynaecologists	Question 1	We note your QS are not pregnancy specific. You may wish to consider making reference to the relevant RCOG Guideline which addresses Sickle cell disease (and acute painful crisis) in pregnancy.
14	Royal College of Obstetricians and Gynaecologists	Question 1	There should be some awareness of not using NSAIDs in pregnancy especially after 28 weeks
15	The Sickle Cell Society	Question 1	The draft quality standard covers many points that patients are concerned with but there a number of points that patients raised with the Society that aren't covered. These are discussed in following sections of this form The data for the proposed quality measures can be collected and should in order to ensure the effectiveness of the standards is being evaluated
16	The Sickle Cell Society	Question 1	The Sickle Cell Society welcomes the opportunity to respond to the NICE quality standards on sickle cell crisis. Our response follows discussions with patients and stakeholders of the Society who attended a patient and parent education day on 19 October 2013. Our response is therefore focused around a number of patient feedback issues, which are outlined in this document. Key Points The standards should include guidelines on providing patients with quick and effective pain relief in sickle cell crisis The standards should extend to ambulance services and primary and community healthcare staff and not just to healthcare professionals in acute settings A clear process or care pathway should be in place for care of sickle cell crisis from ambulance to post discharge from hospital Identification of patients with sickle cell should be easier so patients do not have to prove they suffer from sickle cell disorder when they are admitted to hospital Expertise and specialist centres are important so patients get the best care possible. Currently it is still possible to be admitted to hospital accident and emergency departments, where healthcare professionals in that department are not aware of sickle cell and/or do not have protocols/guidance for the treatment of sickle cell, despite NICE guidance

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17	The Sickle Cell Society	Question 1	Standards for Ambulance Service The standards should extend to ambulance services so that ambulance staff follow set procedures for management of sickle cell crisis. Ambulance staff should be able to ask the right questions to identify a sickle cell crisis and therefore provide pain relief as soon as they can. Ambulance crews should be trained to administer pain relief, and this is even more important with the closure of A&E departments in hospitals in certain areas, leading to longer travel times to get to an accident & emergency.
18	The Sickle Cell Society	Question 1	Evaluation of Guidelines in Practice Evaluating the effectiveness of how the standards are followed is important for improving patient care. If guidelines aren't followed, there should be a thorough review and a penalty imposed. West Midlands Quality Review Service published in September 2013 should form the basis to improve quality standards and should be consulted to ensure patients get the best support in sickle cell crisis.
19	The Sickle Cell Society	Question 1	Community Care Patients noted that at the time of discharge from hospital, no provision is made for those who do not have anyone to take care of them at home whilst they are still recovering. Having some sort of community care support and prevention focus can help with this, where for example community nurses can provide social care services until the patient has recovered. This will lead to lower hospital return episodes. Community care can also provide support to patients in their home when a crisis occurs, where community nurses can carry out blood tests, check oxygen levels and administer pain relief at home rather than patients having to go into hospital. This will make patients feel more at ease and will help hospitals who are struggling to accommodate high patient numbers.
20	Faculty of Pain Medicine	Question 1 and 2	We find that the draft quality standard accurately reflects the key areas for quality improvement and that it would be possible to collect audit data for most of the proposed quality measures. It was suggested to use the NICE clinical guideline 143 clinical audit tool for this purpose. The clinical audit tool would require revision to achieve this, we make comments on specific points in the appropriate section.
21	Faculty of Pain Medicine	Quality Statement 1	No changes to the draft quality standard document proposed. The clinical audit tool, Primary Analgesia Q9 (pethidine), must allow exceptions for patients who are allergic to other opioids.
22	Royal Pharmaceutical Society	Quality Statement 1	The timely provision of appropriate analgesia should be clearly identified as a multidisciplinary issue within the standards and the process of data collection should reflect the different aspects of care that could impact on the timeliness in treatment e.g prescribing delays etc. Pharmacists as part of the multi-disciplinary team have an important role in ensuring that adequate supply systems are in place to ensure clinical areas have access to medicines needed routinely as well as providing other services which would support this quality standard such as medicines reconciliation and providing advice on choice and use of medicines.
23	The Sickle Cell Society	Quality Statement 1	Timely Pain Relief and Treatment The new standards should outline key steps to identify quickly and efficiently what pain relief each patient in a sickle cell crisis requires. Delays in administering analgesia should be minimal and any related assessment should be

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			undertaken quickly so patients are not left in pain for a long period of time. The standards should also outline the treatment of pain for the ambulance service, effectively improving pain relief management in a sickle cell crisis. All possible treatments should be discussed with patients, including Red Cell Exchange, which isn't set out in the current guidelines. It is not clear to patients why this is the case because feedback from patients and carers suggest this option is quicker, effective and more comfortable than manual exchange.
24	Faculty of Pain Medicine	Quality Statement 2	In our clinical experience, reassessment every 30min is difficult to achieve beyond the initial 1-2 hours. The suggested timescale for pain assessment is incongruent to the assessment for opioid-induced adverse effects (every hour) which may lead to inappropriate opioid prescribing or overreliance on intravenous (as supposed to transmucosal, oral or subcutaneous) opioids. We acknowledge that the NICE clinical guideline 143 had reached consensus on this recommendation and agree with their introduction statement "The primary goal in the management of an acute painful sickle cell episode is to achieve effective pain control both promptly and safely." As it would be impossible to audit the achievement of "satisfactory pain relief" in the absence of a definition we propose to formulate the quality standard as "assessment of pain relief every 30min for the first hour after presentation to hospital", and to align the timescale thereafter with the assessment for adverse effects: hourly for the first 6 hours and then at least every 4 hours.
25	Association of Paediatric Anaesthetists of Great Britain and Ireland	Quality Statement 3	Individual drugs are not mentioned in the quality standard, however the "weaker" opioid, Codeine has historically been a treatment option. Given the recent publication from the MHRA, and the joint statement from the RCPCH, APA and RCoA and the genetic predisposition of the Afro-Caribbean population to be ultra-fast metabolizers (CYP2D6) the usefulness of codeine in this population is less clear. Whatever the answer this emphasises the need for continuous SaO2 monitoring and hourly observations for those patients on "weaker" such as codeine, as well as "strong" opioids, and consideration given to this being included in the quality standard.
26	Faculty of Pain Medicine	Quality Statement 3	We suggest removing "blood pressure" from the list of required regular reassessments; it should be listed as "if clinically indicated". Repeated blood pressure measurements can lead to sickling in the affected arm and are often the reason why sickle cell patients might refuse to cooperate with clinical observations. Reduced sedation score and oxygen saturation (measured without nasal or mask oxygen) are the most important indicators for opioid-induced respiratory depression; the latter also aids early detection of acute chest syndrome. It should be highlighted that monitoring must restart at hourly intervals when analgesia is stepped up, e.g. commencement of modified release strong opioids or intravenous PCA. Temperature is only required every 4 hours.
27	Association of Paediatric Anaesthetists of Great Britain and Ireland	Quality statement 4	Whenever SaO2 is measured the FiO2 (inspired oxygen) needs to be recorded, SaO2 recorded as a number in isolation has little relevance. This is particularly important when there are complications of the disease, Acute Chest Syndrome, or of the analgesia provided.
28	Faculty of Pain Medicine	Quality Statement 4	To ensure awareness of healthcare professionals that acute chest syndrome is a potential complication is an important quality measure, in our view it is most strongly linked to training. Patient representatives highlighted during the consultation on the NICE clinical guideline that they see a need for quality improvement in training of health professionals, an area that the draft quality standard document addressed in its introduction (page 3). However it is

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			not clear how this measure can be audited and what data should be collected. Further guidance is recommended, e.g. mandatory training for staff of emergency departments and acute medicine departments in high prevalence areas. An alternative would be measurement of outcome quality: staff awareness about acute chest syndrome being a possible complication (see comment on quality statement 5 below).
29	Faculty of Pain Medicine	Quality Statement 4	The audit of process quality for this measure will rely on the documentation of the denominating conditions: e.g. abnormal respiratory signs and symptoms. Data collection for this audit may be difficult or unreliable as such conditions may be documented in various documents or formats. The use of a formal checklist for acute chest syndrome may be suggested.
30	Faculty of Pain Medicine	Quality Statement 4 (and 5)	This is the only quality statement (5) containing measurement of outcome, namely staff knowledge/awareness on how to access local protocols. During such audit it may be possible to measure outcome for quality statement 4: staff awareness about acute chest syndrome being a possible complication.
31	British Pain Society	Quality Statement 5	The range of specialist support should include specific reference to the availability of access to multidisciplinary pain clinics. This is because this group of patients are vulnerable to complications of long-term or repeated use of strong opioids and if there are concerns or difficulties with reducing or maintaining effective doses then expertise should be readily available. This will be most easily accessed through established pain clinics.
32	Royal Pharmaceutical Society	Quality Statement 5	The production and review of locally agreed protocols on treatment and management of acute sickle cell disease should also be identified as a multidisciplinary issue with the appropriate support from different healthcare professionals including pharmacists being made available. As experts in medicines, pharmacists provide advice on how to take medicines, adverse effects, possible interactions and cautions, to raise patients' awareness and increase their understanding of their condition and therapy and would therefore be ideally placed to form part of these teams.
33	The Sickle Cell Society	Quality Statement 5	Identification of Patients with Sickle Cell The standards should help healthcare professionals correctly identify the acute pain of sickle cell crisis so patients do not have to prove they are suffering, as is often the experience of individuals with sickle cell and their families. Patients felt that hospitals and ambulance put the onus on the patient to prove they are suffering from a crisis, which inevitably causes a delay in treatment and pain management. The use of resources such as identification cards, databases and registers can help identify those with sickle cell in an effective way. Easy identification of sickle cell disorder in patients will help with following protocol without delay. This process can be linked to the National Haemoglobinopathy Registry, which the hospitals should populate and use as a reference.
34	The Sickle Cell Society	Quality Statement 5	Expertise and Specialist Centres A key suggestion for the guidelines is for medical staff and hospitals to gain more knowledge and expertise on sickle cell, which will make them better equipped to provide the best patient care during a sickle cell crisis. Regular training on Sickle Cell Disorder will facilitate this and in turn lead to better patient care. Patient needs can be better assessed and hospitals can develop and aspire to specialist centre status. This will help ambulance crews identify which hospital is better suited for the patient and the patient will in turn not be faced with long periods of waiting in

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			pain. Patients also highlighted that they should be able to go to a preferred hospital rather than the closest or most convenient, especially if there are specialised centres. Some patients become familiar with certain hospitals where staff know their individual care plan or protocol, in turn making the patient comfortable they will get the most consistent and suitable treatment for the individual. The recent peer review of adult haemoglobin services undertaken by the West Midlands Quality Review Service published in September 2013 highlights this point. Of concern to patients, the report also shows worrying variability of the quality of services and thus the patient experience and access. As the only national charity for Sickle Cell Disorder, the Sickle Cell Society is well placed to provide training and support through advocacy and information for better quality care and services for patients and healthcare professionals