NATIONAL INSTITUTE FOR HEALTH AND CLINICAL EXCELLENCE

SCOPE

1 Guideline title

Sickle cell acute episode: management of an acute painful sickle cell episode in hospital

1.1 Short title

Sickle cell acute episode

2 The remit

The Department of Health has asked NICE: 'To produce a clinical guideline on the management of sickle cell crisis in hospital.'

The scope refers to sickle cell crisis as an acute painful sickle cell episode.

3 Clinical need for the guideline

3.1 Epidemiology

a) Sickle cell disease (SCD) 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 racial groups. Sickle cell disease can have a significant impact on morbidity and mortality, although life expectancy has improved in recent years as a result of improvements in patient care and treatment options.

b) Acute painful sickle cell episodes are caused by the sickling process, in which the red blood cells change shape and lead to the blockage of small vessels and tissue infarction. Crises are often

unpredictable and pain may vary in intensity. Repeated crises may result in organ damage.

c) It is estimated that there are between 12,500 and 15,000 people with sickle cell disease in the UK. The prevalence of disease is increasing because of immigration into the UK. The National Sickle Cell and Thalassaemia newborn screening programme also means that more cases are being diagnosed.

d) The distribution of disease reflects that of the multiracial population in the UK: about two thirds of people with sickle cell disease live in London, with the majority of others in major urban areas such as the West Midlands and Manchester. The geographical distribution of sickle cell disease is widening through immigration into other parts of the UK and the increasing mobility of the population.

3.2 Current practice

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

b) The approach to pain management follows the WHO stepladder of non-opioid and opioid analgesia. Treatment begins with a simple oral analgesic in conjunction with non steroidal anti-inflammatory drugs (NSAIDs) and progresses though to weak then stronger opiates, such as morphine and diamorphine.

c) Guidance from the British Committee for Standards in Haematology (2003) recommends that:

- Analgesia should be given within 30 minutes of entering the hospital and effective analgesia should be achieved by 60 minutes.

- Pain, respiratory rate and sedation should be assessed every 20 minutes until pain is controlled.
- A multidisciplinary approach should be used, involving haematologists, paediatricians (for children), pain teams, anaesthetists, psychologists, physiotherapists and counsellors.
- Paracetamol and NSAIDs should be used with opioids to control severe pain.
- Pethidine should not be used for routine analgesia.
- Morphine or diamorphine should be used as first-choice opiate analgesics.
- In children, oral analgesia should be used if possible, although very severe pain may require initial control with pareneteral analgesia.
- Patients should be monitored every 2 hours for pain control (using a pain chart), sedation, respiratory rate and oxygen saturation, and every 4 hours for temperature and pulse.

d) The Sickle Cell Society's standards for clinical care recommend that:

- People presenting with acute sickle pain should receive a first dose of effective analgesia within 30 minutes of arrival, with the aim that pain should be controlled within 2 hours. Pain and sedation scores should be recorded systematically and treatment adjusted accordingly.
- On admission, patients should be assessed for acute and potentially life-threatening complications, including infection, acute chest syndrome, neurological problems, acute renal failure and priapism. Observations for such complications should continue to be recorded regularly, and acted on, throughout every episode of care.

e) .

4 The guideline

The guideline development process is described in detail on the NICE website (see section 6, 'Further information').

This scope defines what the guideline will (and will not) examine, and what the guideline developers will consider. The scope is based on the referral from the Department of Health.

The areas that will be addressed by the guideline are described in the following sections. The guideline will cover management from the point at which it is suspected that the patient is having an acute painful sickle cell episode until the pain is under control. This will be separated into defined time periods as appropriate.

4.1 Population

4.1.1 Groups that will be covered

a) Adults, children and young people with diagnosed sickle cell disease who present with an acute painful sickle cell episode.

b) Within this population, consideration will be given to the specific needs of:

- pregnant women, and
- age-specific sub groups.

4.1.2 Groups that will not be covered

a) People who are sickle cell carriers.

b) People who present with a crisis that is not associated with an acute painful sickle cell episode.

4.2 Healthcare setting

a) Secondary and tertiary care in the NHS.

4.3 Clinical management

4.3.1 Key clinical issues that will be covered

a) Inpatient pharmacological management of acute pain with all types of analgesia, including NSAIDs, non-opioids, weak opioids and strong opioids. Note that guideline recommendations will normally fall within licensed indications; exceptionally, and only if clearly supported by evidence, use outside a licensed indication may be recommended. The guideline will assume that prescribers will use a drug's summary of product characteristics to inform their decisions for individual patients.

b) Choice, timing and route of analgesia, including patient controlled analgesia.

c) Timing and frequency of inpatient pain and physiological assessment for monitoring purposes.

d) Adverse events associated with pain management.

e) Inpatient non-pharmacological management of acute pain using:

- distraction techniques
- acupuncture
- transcutaneous electrical nerve stimulation (TENs)
- heat therapy.

f) Clinical signs and symptoms to identify patients who are likely to have acute complications associated with a painful sickle cell episode.

g) Optimal clinical setting for managing episodes of acute pain.

h) Skills of healthcare professionals and teams providing care.

i) The specific information and support needs of adults and children and young people with an acute painful sickle cell episode, and their parents/carers and families, related to pain management.

4.3.2 Clinical issues that will not be covered

a) Managing chronic pain.

b) Preventing an acute painful sickle cell episode.

c) Formal diagnostic investigations to confirm acute complications.

d) Managing acute complications.

e) Managing side effects associated with pharmacological interventions.

f) Sickle cell episodes not associated with acute pain.

g) Co-medications, unless they are used to manage acute pain.

4.4 Main outcomes

a) Survival.

b) Intensity and duration of pain using validated and age-appropriate pain rating scales (this will include parental and healthcare professional assessment for children).

c) Adverse events associated with pain management.

d) Risk factors associated with the development of acute complications.

e) Patient satisfaction or experience of pain management.

f) Health-related quality of life.

g) Resource use and cost.

4.5 Economic aspects

Developers will take into account both clinical and cost effectiveness when making recommendations involving a choice between alternative interventions. A review of the economic evidence will be conducted and analyses will be carried out as appropriate. The preferred unit of effectiveness is the quality-adjusted life year (QALY), and the costs considered will usually only be from an NHS and personal social services (PSS) perspective. Further detail on the methods can be found in 'The guidelines manual' (see ‘Further information’).

The key health economic question for this guideline appears to be the cost effectiveness of different pharmacological strategies for managing an acute painful sickle cell episode. The strategies evaluated will depend on the availability of data on which to base estimates of costs and effects.

Further cost effectiveness analysis will be considered if any additional questions are identified during guideline development.

4.6 Status

4.6.1 Scope

This is the consultation draft of the scope. The consultation dates are 7 June to 5 July 2011.

4.6.2 Timing

The development of the guideline recommendations will begin in August 2011.

5 Related NICE guidance

5.1 Published guidance

- Antenatal care. NICE clinical guideline 62 (2008). Available from www.nice.org.uk/guidance/CG62
- Intrapartum care. NICE clinical guideline 55 (2007). Available from www.nice.org.uk/guidance/CG55

- Acutely ill patients in hospital. NICE clinical guideline 50 (2007). Available from www.nice.org.uk/guidance/CG50
- Preoperative tests. NICE clinical guideline 3 (2003). Available from www.nice.org.uk/guidance/CG3

5.2 Guidance under development

NICE is currently developing the following related guidance (details available from the NICE website).

- Opioids in palliative care. NICE clinical guideline. Publication date to be confirmed.

6 Further information

Information on the guideline development process is provided in:

- 'How NICE clinical guidelines are developed: an overview for stakeholders' the public and the NHS'
- 'The guidelines manual'.

These are available from the NICE website (www.nice.org.uk/guidelinesmanual). Information on the progress of the guideline will also be available from the NICE website (www.nice.org.uk).