NATIONAL INSTITUTE FOR HEALTH AND CLINICAL EXCELLENCE

SCOPE

1 Guideline title

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

1.1 Short title

Sickle cell acute painful 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 ethnic groups. Sickle cell disease can have a significant impact on morbidity and mortality.

b) Acute painful sickle cell episodes are caused by the sickling process. The red blood cells in people with sickle cell disease behave differently under a variety of conditions, including dehydration, low oxygen and elevated temperature. Changes in any of these conditions may cause them to block small vessels and cause tissue infarction. Crises are often unpredictable and pain

may vary in intensity but can be excruciating. 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 National Haemoglobinopathy Registry aims to improve patient care and will provide more accurate information on the number and geographical distribution of patients in the future. The prevalence of the disease is increasing because of immigration into the UK and new births. 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 multi-ethnic 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 non-opioids such as paracetamol and progresses though to weak opioids such as codeine and then stronger opioids such as morphine until the pain is controlled.

c) There is guidance available from the British Committee for Standards in Haematology (2003) and Sickle Cell Society (2008)

relating to the management of acute pain. There is also a guideline for clinical care in children published by the NHS Sickle Cell and Thalassaemia Screening Programme and The Sickle Cell Society (2010). Recommendations from these guidelines cover the type and timing of analgesia, assessment and monitoring of pain and other physiological measures, and the teams involved in caring for patients with an acute painful sickle cell episode.

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 any genotype for 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 subgroups.

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 (such as aplastic crisis).

4.2 Healthcare setting

a) In-hospital settings and specialist centres in the NHS.

4.3 Clinical management

4.3.1 Key clinical issues that will be covered

a) Pharmacological interventions that are used to manage acute painful episodes in hospital. This includes all types of analgesia, including NSAIDs, non-opioids, weak opioids and strong opioids. This also includes oxygen, nitrous oxide and prescribed fluids. 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) When and how often pain and physiological assessments are carried out for monitoring purposes.

d) Non-pharmacological interventions that are used to manage acute painful episodes in hospital.

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

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

g) Skills and knowledge of healthcare professionals and teams providing care.

h) 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, in relation 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 interventions used to manage acute pain.

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) Rates of adverse events that are associated with interventions to manage acute painful episodes in hospital.

d) Development of acute complications.

e) Patient and carer 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 final scope.

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

- Depression in adults with a chronic physical health problem. NICE clinical guideline 91 (2009). Available from www.nice.org.uk/guidance/CG91

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).