NICE National Institute for Health and Care Excellence

2023 exceptional surveillance of sickle cell disease: managing acute painful episodes in hospital (NICE guideline CG143)

Surveillance report
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Surveillance decision

We will not update the NICE guideline on sickle cell disease.

Reasons for the decision

This review considered new evidence on pharmacological interventions for managing an acute painful sickle cell episode (vaso-occlusive crises) in children, young people and adults in a hospital setting. There was insufficient evidence on licensed medicines for this indication to support an update of the recommendations.

For further details and a summary of all evidence identified in surveillance, see appendix A.

Exceptional surveillance review summary

A pain crisis is the most common problem of sickle cell disease and can require several treatments at once, usually in an emergency. The priority is to control the pain, this can be done using medicines (such as opioids, non-steroidal anti-inflammatories and paracetamol).

Reason for the exceptional review

NICE was contacted by NHS England and asked to check if the evidence-base of analgesia for managing an acute painful sickle cell episode had developed sufficiently to support an update to the NICE guideline on sickle cell disease.

Methods

The exceptional surveillance process consisted of:

- Focused literature searches on evidence related to primary analgesia and reassessment and ongoing management.
- Considering new or updated Cochrane reviews.

- Considering relevant information from previous surveillance reviews of the guideline in 2016.
- Examining related NICE guidance and quality standards.
- Examining the NICE event tracker for relevant ongoing and published events.
- A search for ongoing research.
- Assessing the new evidence against current recommendations to determine whether or not to update sections of the guideline.

For further details about the process and the possible update decisions that are available, see ensuring that published guidelines are current and accurate in developing NICE guidelines: the manual.

Search and selection strategy

We searched for new evidence that relates to primary analgesia (including timing) for acute painful sickle cell episodes in hospital, which is covered by 3 areas of the guideline: individualised assessment at presentation, primary analgesia and reassessment and ongoing management.

We found 11 studies in a search for randomised controlled trials (RCTs) and systematic reviews published between 16 February 2016 and 3 February 2023. This included 6 RCTs and 5 systematic reviews. The systematic reviews were all limited by a small number of included studies. There was also heterogeneity across the available evidence base with respect to populations, interventions and settings. Additionally, most studies were small and none of the RCTs were from UK settings.

See appendix A for details of all evidence considered and references.

Selecting relevant studies

Studies were selected in accordance with criteria used for the guideline. In addition, we considered unlicensed drugs (such as ketamine and fentanyl).

Ongoing research

We checked for relevant ongoing research; of the ongoing studies identified, 2 were assessed as relevant to the recommendations. Therefore, we will monitor these ongoing studies and will evaluate the impact of the results on current recommendations as soon as the studies have published the findings. These studies are:

- Hyperbaric-oxygen therapy (HBOT) versus placebo for treating vaso-occlusive crisis in sickle cell disease.
- Nebulized sub-dissociative dose ketamine at three different dosing regimens for treating acute pain in the pediatric emergency department.

See ensuring that published guidelines are current and accurate in developing NICE guidelines: the manual for more details on our consultation processes.

Other relevant NICE guidance

The following NICE technology appraisal guidance is available:

- Crizanlizumab for preventing sickle cell crises in sickle cell disease.

The following guidance topics are in development:

- Voxelotor for treating sickle cell disease (expected publication 23 August 2023).
- Exagamglogene autotemcel for treating severe sickle cell disease (expected publication TBC).

Equalities

People with sickle cell disease face health inequalities because the condition is not well understood, results in disability, and is more common in people of African or African-Caribbean family origin, who tend to also have poorer health outcomes than other ethnicities.

As part of the intelligence gathering for this review we identified the recent All Party Parliamentary Group (APPG) enquiry into avoidable deaths and failures of care for sickle cell patients in secondary care, No one's listening (2021).

The APPG report highlights a distinct lack of investment in sickle cell research and medicines. The current review has sought to identify current and ongoing research within the scope of the surveillance check, as outlined above.

An equalities and health inequalities assessment was completed during this surveillance review. See appendix B for details.

Overall decision

After considering all evidence and other intelligence and the impact on current recommendations, we decided that no update is necessary at this time. However, we will continue monitoring the ongoing studies.

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