

MANAGEMENT OF SICKLE CELL DISEASE

Supporting information

This guideline has been prepared with reference to the following:

Qureshi A, Kaya B, Pancham S et al. Guidelines for the use of hydroxycarbamide in children and adults with sickle cell disease: A British Society for Haematology Guideline. Br J Haematol. 2018;181:460-75

<https://b-s-h.org.uk/guidelines/guidelines/guidelines-for-the-use-of-hydroxycarbamide-in-children-and-adults-with-sickle-cell-disease>

NICE. Sickle cell acute painful episode: management of an acute painful sickle cell episode in hospital. 2012. London. NICE

<https://www.nice.org.uk/guidance/cg143>

Pethidine should be avoided as a means of pain relief?

Pethidine has been associated with fitting in patients with sickle cell disease (Davies, 1997), and for this reason is not recommended for routine use by the British Association for Accident and Emergency Medicine (Tachakra, 1998).

Davies SC, Oni L. Management of patients with sickle cell disease. BMJ 1997;315:656-60
<http://www.bmj.com/content/315/7109/656>

Tachakra SS, Davies SC. Management of sickle cell crisis: British Association for Accident and Emergency Medicine. Guidelines for the management of sickle cell crises. J Accid Emerg Med 1998;15:356-7
<http://emj.bmj.com/content/15/5/356.full.pdf>

Evidence Level: V

Exchange transfusion should be considered in patients with acute chest syndrome and with Hypoxia?

A 2022 review identified the following indications for exchange transfusion:

- Severe hypoxemia (oxygen saturation < 85% or requiring at least 2–4 L supplemental oxygen)
- Multilobar lung disease
- Rapidly progressive ACS or multiorgan injury
- Failing to improve or disease progression following simple transfusion
- High risk of adverse outcomes including prior history of severe ACS, cardiopulmonary disease, or pregnancy (Koehl, 2022)

A systematic review (Dolatkah, 2020) failed to find any RCTs which showed how effective blood transfusions might be for treating acute chest syndrome in people with sickle cell disease.

Dolatkah R, Dastgiri S. Blood transfusions for treating acute chest syndrome in people with sickle cell disease. Cochrane Database Syst Rev. 2020:CD007843
<https://www.cochranelibrary.com/cdsr/doi/10.1002/14651858.CD007843.pub4/full>

Koehl JL, Koyfman A, Hayes BD et al. High risk and low prevalence diseases: Acute chest syndrome in sickle cell disease. Am J Emerg Med. 2022;58:235-44

Evidence Level: III

Last amended March 2023
Last reviewed January 2026