

Sickle Cell Disease

EVIDENCE REPOSITORIES

Evidence repositories are collections of best available resources and evidence (clinical guidelines, peer reviewed literature, systematic reviews, etc.), collated by our knowledge synthesis team and content advisors. This evidence repository is not intended to be an exhaustive list of resources for a topic, but rather a curated list of current, evidence-based resources, based on expert consensus of relevance and usability for a general emergency department setting. We search databases (Cochrane Library, PubMed, TRIP Database) and web search engines (Google, Google Scholar) to locate evidence. Additionally, hospital websites are browsed for guidance documents, such as clinical practice guidelines (CPG) for healthcare professionals

Every effort is made to identify resources that are open access (i.e. publicly available, free of charge, not requiring a subscription).

More information about the creation of our evidence repositories can be found at <https://pubmed.ncbi.nlm.nih.gov/28537762/>

CONTENT TEAM

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Key Resources

1. U.S. Department of Health and Human Services, Centers for Disease Control. [3 Tips about sickle cell disease every emergency provider needs to know](#). 2022.
2. Emergency Medicine Residents' Association. [Overview of complications in pediatric patients with sickle cell disease and how to manage them in the emergency department](#). 2021.
3. American College of Emergency Physicians. [Managing sickle cell disease in the ED](#). 2022.
4. Centers for Disease Control and Prevention. [Sickle cell disease](#). 2022.
5. Beck CE, Trottier ED, Kirby-Allen M, Pastore Y, Canadian Paediatric Society Acute Care Committee. [Acute complications in children with sickle cell disease: Prevention and management](#). *Paediatr Child Health*. 2022;27(1):50-55.

Clinical Guidelines

1. US Department of Health and Human Services. [Evidence-based management of sickle cell disease expert panel report, 2014: Guide to recommendations](#). 2014.
2. Brandow AM, Carroll PC, Creary S, et al. [American Society of Hematology 2020 guidelines for sickle cell disease: management of acute and chronic pain](#). *Blood Adv*. 2020;4(12):2656-2701.
3. DeBaun MR, Jordan LC, King AA, et al. [American Society of Hematology 2020 guidelines for sickle cell disease: Prevention, diagnosis, and treatment of cerebrovascular disease in children and adults](#). *Blood Adv*. 2020;4(8):1554-1588.
4. Canadian Haemoglobinopathy Association. [Consensus statement on the care of Patients with sickle cell disease in Canada](#). 2014.
5. U.S. Department of Health and Human Services. [The management of sickle cell disease](#). 2002.
6. Children's Health Queensland Hospital and Health Services. [Sickle Cell Crisis: Emergency Management in Children](#). 2018.
7. The Royal Children's Hospital Melbourne. [Clinical Practice Guidelines: Sickle cell disease](#). 2022.

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Systematic Reviews

1. Yawn BP, Buchanan GR, Afenyi-Annan AN, et al. Management of sickle cell disease: Summary of the 2014 evidence-based report by expert panel members [published correction appears in JAMA. 2014 Nov 12;312(18):1932] [published correction appears in JAMA. 2015 Feb 17;313(7):729]. *JAMA*. 2014;312(10):1033-1048.
2. Harris EM, Vilk E, Heeney MM, et al. A systematic review of ketamine for the management of vaso-occlusive pain in sickle cell disease. *Pediatr Blood Cancer*. 2021;68(7):e28989.
3. Subramaniam S, Chao JH. Managing Acute Complications Of Sickle Cell Disease In Pediatric Patients. *Pediatr Emerg Med Pract*. 2016;13(11):1-28.

Key Studies

1. Brousseau DC, MD, MS, Alpern ER, Chamberlain JM, et al. A multiyear cross-sectional study of guideline adherence for the timeliness of opioid administration in children with sickle cell pain crisis. *Annals of Emergency Medicine*. 2020;76(3).
2. Lin SM, Strouse JJ, Whiteman LN, Anders J, Stewart RW. Improving quality of care for sickle cell patients in the pediatric emergency department. *Pediatr Emerg Care*. 2016;32(1):14-16.
3. Corriveau-Bourque C, Bruce AA. The Changing Epidemiology of Pediatric Hemoglobinopathy Patients in Northern Alberta, Canada. *J Pediatr Hematol Oncol*. 2015;37(8):595-599.
4. Paquin H, Trottier ED, Pastore Y, Robitaille N, Dore Bergeron MJ, Bailey B. Evaluation of a clinical protocol using intranasal fentanyl for treatment of vaso-occlusive crisis in sickle cell patients in the emergency department. *Paediatr Child Health*. 2020;25(5):293-299.
5. Paquin H, D Trottier E, Robitaille N, Pastore Y, Dore Bergeron MJ, Bailey B. Oral morphine protocol evaluation for the treatment of vaso-occlusive crisis in paediatric sickle cell patients. *Paediatr Child Health*. 2019;24(1):e45-e50.
6. Glassberg JA. Improving emergency department-based care of sickle cell pain. *American Society of Hematology*. 2017;412-417.