CHOC Children's
Nursing Research

CHOC Children's Hospital
Best Evidence and Recommendations

Best Practice for Sickle Cell Pain Crisis

Emily Gruendyke, MSN, RN, CPHON
egruendyke@choc.org

PICO: In pediatric sickle cell patients in pain crisis, how does current institutional management compare to evidence-based practice guidelines as presented by the National Heart, Lung, and Blood Institute (NHLBI) regarding cost, time to early intervention pain management, admission rates, and length of stay?

P (Population/problem): In pediatric sickle cell patients in pain crisis
I (Intervention/issue): how does current institutional management
C (Comparison): compare to evidence-based practice guidelines as presented by NHLBI
O (Outcome): cost, time to early intervention pain management, admission rates, and length of stay?

Background:

Sickle Cell Disease (SCD) is the most common genetic blood disorder in the United States, affecting approximately 100,000 Americans (CDC, 2017). Due to the crescent shape of the sickled red blood cells, clumping of the cells in the vasculature can lead to a highly painful ischemic process which is caused by the decreased circulation of oxygen to the tissue (U.S. Department of Health and Human Services, 2014). These highly painful episodes in SCD patients are known as "pain crises" or "veno-occlusive crises." A pain crisis is an emergency for the patient and requires aggressive pain medication administration, which often includes intravenous opioids and may lead to hospitalization.

Pain is the most common complication of SCD and is the leading cause of emergency department visits and hospitalizations in the pediatric population (Brousseau, Owens, Mosso, Panepinto, & Steiner, 2010). Not only are children hospitalized with SCD pain crisis missing school and social events, but a high financial cost is incurred for each hospitalization. In a study by Bou-Maroun and colleagues (2018), 75,234 pediatric SCD inpatient hospitalizations were reviewed. More than 64% of those were for pain crises and collectively had an average annual expenditure of $588,632,958.

The NHLBI has published SCD care guidelines with evidence-based recommendations for acute pain crises in sickle cell disease. NHLBI (2014) recommendations include the administration of analgesics within 30 minutes of triage and the use of an individualized prescribing/SCD-specific protocol to help facilitate rapid, effective, and safe medication administration. The literature search conducted for this project specifically looked for studies that implemented strategies recommended by the NHLBI.

Search Strategies and Databases Reviewed:

- Databases searched for this review included CINAHL, Medline in EBSCO, and PubMed. Key search words: sickle cell pain crisis, veno-occlusive crisis, emergency department, pediatric, admission rate, length of stay, NHLBI. This search yielded nine relevant articles, and five of these articles had the most substantial and compelling results.

- Websites reviewed included the NHLBI website and the SCD pain algorithms on the websites of Texas Children's Hospital and Children's Hospital of Philadelphia.
- A request for institutional SCD pain algorithms was posted in the online forum for the Association of Pediatric Hematology Oncology Nurses, and two respondents attached the algorithms.
- Emails were sent to two leading children's hospitals on the West Coast and one leading children's hospital on the East Coast. It was found that the West Coast hospitals did not have protocols for SCD pain.

Synthesis of Evidence:

- Clinical pathways improve the time to the first administered analgesic and opioid (Ender et al., 2014).
- Earlier IV pain medication administration leads to a shorter emergency department length of stay, though it may not lead to a shorter length of stay for admitted patients (Lin, Strouse, Whiteman, Anders, & Stewart, 2016).
- Individualized pain plans decrease admission rates and increase patient satisfaction (Krishnamurti et al., 2014).
- Standardized, time-specific SCD pain protocols increase the occurrence of the first parenteral pain medication being administered within 30 minutes of triage (Kavanagh et al., 2015).
- Earlier achieved maximum opioid dose is linked to a shorter length of stay (Payne et al., 2018).
- Individualized pain plans (IPP) can decrease admission rates and hospital costs/patient charges. Receiving a second opiate dose within 45 minutes of the first dose also can lower the admission rate (Schefft, Swaffar, Newlin, Noda, & Sisler, 2018).

Practice Recommendations:

- Develop an SCD pain care guideline and order set for the acute care unit and the emergency department.
- Develop an individualized SCD pain plan that would be completed on all children during any inpatient or outpatient visit and then made easily accessible within the electronic medical record.
- Educate providers and nurses in Emergency Department and Hematology about SCD and the care guideline before implementation. Include instructions on completing an individualized SCD pain plan and accessing the plan in the electronic medical record.
- Complete a satisfaction survey of patients and families before and after implementation of the individualized SCD pain plan to assess effectiveness.

Acknowledgements:

- The Evidence-Based Scholars Program was supported by a grant from the Walden W. and Jean Young Shaw Foundation
- Jennifer Hayakawa, DNP, PCNS-BC, CNRN, CCRN, Nurse Scientist, CHOC Children's.
- Vicky R. Bowden, DNSc, RN, Azusa Pacific University, CHOC Children's Hospital EBP Scholars Mentor.

References:

Bou-Maroun, L. M., Meta, F., Hanba, C. J., Campbell, A. D., & Yanik, G. A. (2018). An analysis of inpatient pediatric sickle cell disease: Incidence, costs, and outcomes. *Pediatric Blood & Cancer*, *65*(1), e26758.

Brousseau, D. C., Owens, P. L., Mosso, A. L., Panepinto, J. A., & Steiner, C. A. (2010). Acute care utilization and rehospitalizations for sickle cell disease. *JAMA*, *303*(13), 1288-1294.

Center for Disease Control (CDC). (2017). *Data and statistics on sickle cell disease.* Retrieved from https://www.cdc.gov/ncbddd/sicklecell/data.html

Ender, K. L., Krajewski, J. A., Babineau, J., Tresgallo, M., Schechter, W., Saroyan, J. M., & Kharbanda, A. (2014). Use of a clinical pathway to improve the acute management of vaso-occlusive crisis pain in pediatric sickle cell disease. *Pediatric Blood & Cancer*, *61*(4), 693-696.

Lin, S. M., Strouse, J. J., Whiteman, L. N., Anders, J., & Stewart, R. W. (2016). Improving the quality of care for sickle cell patients in the pediatric emergency department. *Pediatric Emergency Care*, *32*(1), 14-16

Kavanagh, P. L., Sprinz, P. G., Wolfgang, T. L., Killius, K., Champigny, M., Sobota, A., ... & Moses, J. M. (2015). Improving the management of vaso-occlusive episodes in the pediatric emergency department. *Pediatrics*, *136*(4), e1016-e1025.

Krishnamurti, L., Smith-Packard, B., Gupta, A., Campbell, M., Gunawardena, S., & Saladino, R. (2014). Impact of individualized pain plan on the emergency management of children with sickle cell disease. *Pediatric Blood & Cancer*, *61*(10), 1747-1753.

Melzer-Lange, M. D., Walsh-Kelly, C. M., Lea, G., Hillery, C. A., & Scott, J. P. (2004). Patient controlled analgesia for sickle cell pain crisis in a pediatric emergency department. *Pediatric Emergency Care*, *20*(1), 2-4.

Payne, J., Aban, I., Hilliard, L. M., Madison, J., Bemrich-Stolz, C., Howard, T. H., ... & Lebensburger, J. D. (2018). Impact of early analgesia on hospitalization outcomes for sickle cell pain crisis. *Pediatric Blood & Cancer*, *65*(12), e27420.

Schefft, M. R., Swaffar, C., Newlin, J., Noda, C., & Sisler, I. (2018). A novel approach to reducing

admissions for children with sickle cell disease in pain crisis through individualization and standardization in the emergency department. *Pediatric Blood & Cancer*, *65*(10), e27274.

U.S. Department of Health and Human Services. (2014). *Evidence-based management of sickle cell disease*. Retrieved from https://www.nhlbi.nih.gov/sites/default/files/media/docs/sickle-cell disease-report%20020816_0.pdf