SICKLE CELL PAIN CLINICAL PATHWAY

EXECUTIVE SUMMARY

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Primary Objective

Vaso-Occlusive crisis significantly negatively affects health outcomes and patients' quality of life with SCD. In addition, pain associated with the vaso-occlusive problem has been hard to manage, leading to unnecessary suffering, increased emergency room visits, prolonged hospital admissions, and healthcare costs (Ender et al., 2014). Therefore, there is a need for a standardized treatment pathway when a pediatric patient is in the emergency room experiencing pain related to a vaso-occlusive crisis. The aims of implementing this pathway include:

1. Standardizing sickle cell acute pain treatment by utilizing a sickle cell pain pathway.
2. Improve the time from arrival to the first pain medication administration.
3. Improve the time between subsequent opioid doses.
4. Improve the frequency of pain reassessment after pain medication administration.
5. Improve the percentage of patients who receive two doses of IN fentanyl.

Recommendations

Multi-modal Pain Medicine Administration

Multimodal analgesia includes two or more drugs that provide analgesia via different mechanisms (Kenney & Smith, 2022). Opioids are traditionally the gold standard for treating pain in patients with sickle cell disease using a multimodal analgesia approach; other analgesics would be added in conjunction with the opioid. These analgesics include acetaminophen, nonsteroidal anti-inflammatory drugs, ketamine, lidocaine, etc. (Kenney & Smith, 2022). When utilizing a multimodal analgesic approach, the goal is to use medications with different mechanisms of action, target other pain receptors, and reduce adverse effects (Baichoo, Asuncion, and Chaar, 2019). Outpatient medication management includes a 3-day opioid bridge until they see their sickle cell disease provider. Patients should also be educated on using acetaminophen and oral NSAIDs for pain management (ACEP, 2021).

Opioids

The use of opioids to treat pain in sickle cell patients experiencing a vaso-occlusive crisis is the most common treatment (Arzoun et al., 2022). Opioids have an analgesic effect due to their interaction with opioid receptors (Niscola et al., 2009). The National Heart, Lung, and Blood Institution recommend administering an IV opioid within 60 minutes of arrival to the emergency department (Arzoun et al., 2022). In addition, the American College of Emergency Physicians recommends initiating aggressive pain management using opioids and NSAIDs (Zempsky, 2010). All patients receiving IV opioids should be placed on continuous oxygen monitoring. Pain should be reassessed within 30 minutes of opioid administration, and if the pain is still moderate to severe, a repeat opioid dose should be considered (Zempsky, 2010). Opioids to consider are intranasal fentanyl, IV morphine, and IV hydromorphone.

Ketamine

Ketamine can be utilized to significantly reduce pain and opioid utilization in patients experiencing a sickle cell vaso-occlusive crisis (Alshahrani & Alghamdi, 2021). Ketamine causes an analgesic effect "by blocking N-methyl-D-aspartate (NMDA) receptors, which impairs sensitization of spinal neurons to nociceptive stimuli" (Harris et al., 2022). Ketamine can be used in conjunction with opioids and dexmedetomidine (Froomkin et al., 2022). American Society of Hematology 2020 recommends subanesthetic ketamine infusion as adjunctive treatment to treat vaso-occlusive pain crisis in pediatric patients.

Dexmedetomidine

Dexmedetomidine is an alpha 2 adrenoreceptor agonist that has sedative and analgesic properties (Sheehy et al., 2015). Dexmedetomidine is utilized in conjunction with analgesic therapy to help with vaso- occlusive pain crisis (Sheehy et al., 2015). It has been proven to reduce opioid requirement, help facilitate opioid weaning and decrease pain scores (Sheehy et al., 2015). Dexmedetomidine is typically given at a continuous infusion and titrated as needed for pain management (Sheehy et al., 2015).

Nonsteroidal Anti-Inflammatory Drug

Nonsteroidal anti-inflammatory (NSAID) drugs, such as ibuprofen/ketorolac, have been utilized to treat pain in sickle cell patients. NSAIDs treat pain by inhibiting nonselective cyclooxygenase enzymes and suppressing the production of prostaglandins (Han et al., 2017). It has been found that NSAIDs alone are not effective enough to decrease pain, but when combined with an opioid (Han et al., 2017). American Society of Hematology 2020 recommends a 5–7-day course of NSAIDs and opioids to treat acute pain in patients with sickle cell disease. It is essential to consider the kidney and renal function of the patient when using NSAIDs.

Acetaminophen

Intravenous (IV) acetaminophen was first approved in 2010 for children over the age of two to treat pain (Baichoo, Asuncion, and Chaar, 2019). IV acetaminophen has an analgesic effect beginning at 5-10 minutes, peaking at one hour, and lasting for four to six hours (Baichoo, Asuncion, and Chaar, 2019). A study by Baichoo, Asuncion, and Chaar (2019) examined the effect of IV acetaminophen when treating vaso-occlusive crisis in children with sickle cell pain. In addition, a retrospective study was done to look at pain management in patients with vaso-occlusive crisis, 28 patients received an opioid, and 18 received an opioid and IV acetaminophen (Baichoo, Asuncion, and Chaar, 2019). This study found that in the group of children who received IV acetaminophen and an IV

opioid, there was a 14.6% pain reduction compared to those who only received an opioid (Baichoo, Asuncion, and Chaar, 2019).

Timing of Pain Medicine Administration and Reassessment

American Society of Hematology 2020 found that in adults and children with SCD who present with acute pain, an analgesic should be administered within one hour of arrival, and pain assessments should occur every 30 minutes after pain medication is given (Brandow et al., 2020). A clinical pathway time to opioid administration can be decreased, which ASH supports. Another way that this recommendation could be met is by using intranasal medication when available (Brandow et al., 2020). Pain requires immediate intervention, typically within 60 minutes of arriving at the emergency room (ACEP, 2021). With children experiencing a pain episode, internasal fentanyl should be considered first-line treatment (ACEP, 2021). Opioids can be re-dosed every 30 minutes until the pain improves. Pain reassessments should be 15-30 minutes after administering pain medication (ACEP, 2021). It is recommended that aggressive pain management be initiated by utilizing opioids and NSAIDs (Zempsky, 2010). Pain should be reassessed within 30 minutes of opioid administration, and if the pain is still moderate to severe, a repeat opioid dose should be considered (Zempsky, 2010).

Pain Documentation

Pain in children with sickle cell disease can be hard to assess due to the complexity of the pain that they experience and difficulties understanding and describing their pain (Zempsky, 2010). When evaluating a patient's pain, utilizing the correct pain scale is essential if they cannot self-report their pain (Niscola et al., 2009). Self-reported pain is the gold standard for pain assessment. Frequently utilized pain scales are visual analog, verbal, numerical, and Wong-Baker face scales (Niscola et al., 2009). The pain scale used must be appropriate for the patient's developmental age. The Society of Hematology 2020 recommends pain reassessment every 30-60 minutes to ensure pain is well controlled (Brandow et al., 2020).

Rationale

Sickle cell disease (SCD) is the most common genetic disease in the United States (Sedrak & Kondamudi, 2022). Around 100,000 people live with sickle cell disease in the United States and 20 million worldwide, primarily African American (Sedrak & Kondamudi, 2022). Despite recent advancements in the care of people with sickle cell disease, a study done by Lubeck et al. (2019) found that people living with SCD have a shorter life span, higher morbidity related to side effects, a higher burden of medical costs, and higher loss of income related to the disease process. According to Lubeck et al. (2019), people with SCD spend an average of 5 to 6 days in the hospital per admission, have an average of 2 to 3 hospital admissions per year, and have a total of $700,000 in loss of lifetime income due to their disease. It was also found that SCD causes high morbidity rates, including

pain, vaso-occlusive crisis, fatigue, depression, reduced social and school functioning, and diminished well-being (Lubeck et al., 2019). These morbidities lead to an overall lesser quality of life ratings among those living with SCD (Lubeck et al., 2019). 60% of people with sickle cell disease report having one severe pain episode yearly, leading to emergency room visits or hospitalization (Ender et al., 2014). Vaso-occlusive crisis can lead to unnecessary suffering, increased emergency room visits, prolonged hospitalizations, and early death compared to others without sickle cell disease (Ender et al., 2014). When pain is uncontrolled or not identified, the patient experiences adverse effects, including psychosocial, poorer quality of life, increased distress, and poorer health outcomes (CDC, 2020). The American Society of Hematology, the American College of Emergency Physicians and Clinical Pediatric Emergency Medicine support using a clinical pathway to improve the care of sickle cell patients who present to the emergency department experiencing an acute pain crisis. Pathways have been found to improve patient outcomes, reduce barriers, and deliver safe, high-quality, and effective care. Pathways addressing pain in patients with sickle cell disease should address the number of analgesic doses administered, the time of first analgesic, and the proportion discharged home from ED (Brandow et al., 2020), (ACEP, 2021), (Zempsky, 2010). The best practice for all organizations is to implement a clinical pathway to improve the patient outcomes of sickle cell disease patients.

Metrics

- **Outcome**
  - Decrease the average time from triage time in ED to first IV/IN pain medication
  - Decrease the average time from first IV/IN pain medication to first pain reassessment
  - Decrease the average time from first IV opioid to second IV opioid
  - Increase the percentage of patients in moderate to severe pain who receive two doses of IN fentanyl
- **Process**
  - Increase the proportion of eligible patients who utilize the pathway order set
- **Balancing**
  - Monitor ED length of stay
  - Monitor the proportion of patients admitted from ED
  - Monitor the proportion of patients readmitted within 48 hours for a Sickle Cell Disease Pain Crisis

Team Members

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Evidence

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