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Treatment of Vaso-Occlusive Pain Related to Sickle Cell Disease in Pediatric Patients: A Systematic Review

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Studies were included in the review if they were randomized controlled trials. Trials must include participants 18 years and younger with sickle cell disease. The intervention must be measured in the trial must be reduction in pain score; no standard pain score exists so “opioid”, “non-opioid”, “NSAID”, “pediatric”, and “children”. Sickle cell disease (SCD) is a rare inherited disorder that affects the shape of erythrocytes. SCD is estimated to affect 70,000 - 100,000 Americans and is seen predominately in African-Americans.1 In SCD, the body produces sickle-shaped red blood cells, which have shorter lifespans than normal red blood cells and can lead to anemia. The abnormal shape causes them to become lodged in blood vessels, causing pain and organ damage. Symptoms generally begin by three years of age and complications of sickle cell disease include vaso-occlusive crises, acute chest syndrome, and others.2 A vaso-occlusive crisis is an episode of pain, described as sharp, intense, and throbbing, most commonly occurring in the lower back, leg, hip, abdomen, or chest. It typically begins at night and lasts 3-14 days. It is the most common reason for hospitalization in patients with SCD.3 Episodes increase in frequency throughout childhood.2 No cure for SCD is widely available; treatment is symptomatic and supportive. No evidence based guidelines for the treatment of pain during vaso-occlusive crises in pediatric patients with SCD are published.2

**INTRODUCTION**

Sickle cell crisis (Pain) is the deplorable cause of hospitalization in the people with SCD.4 Episodes increase in frequency throughout childhood.2 No cure for SCD is widely available; treatment is symptomatic and supportive. No evidence based guidelines exist for the treatment of pain during vaso-occlusive crises in pediatric patients with SCD.

**METHODOLOGY**

Studies were included in the review if they were randomized controlled trials. Trials must include participants 18 years and younger with sickle cell disease. The intervention must be a FDA approved medication (not necessarily approved for SCD). One of the outcomes must be reduction in pain score; no standard pain score exists so “opioid”, “non-opioid”, “NSAID”, “pediatric”, and “children”. Literature searches were conducted using PubMed, Embase, Cochrane Central Register, and ClinicalTrials.gov. Search terms included (but were not limited to): “sickle cell disease”, “sickle cell crisis”, “vaso-occlusive crisis”, “pain management”, “analgesia”, “opioid”, “non-opioid”, “NSAID”, “pediatric”, and “children”.

**RESULTS**

A comprehensive literature search provided 1,094 results, including duplicate publications. Screening of the literature search results identified 16 trials that needed further evaluation. Six trials were included in the systematic review, after application of the inclusion and exclusion criteria. Studies are summarized in Figure 3 and Table 1.

**CONCLUSIONS**

Based on the results of the literature review, intranasal fentanyl and L-arginine have some evidence for use in pediatric patients with vaso-occlusive crises. More evidence is needed; however both may be reasonable, safe options in patients with uncontrolled pain as an adjunct to standard therapy.

**REFERENCES**

5 National Initiative on Pain Control. Pain Assessment Scales. URL: http://www.paindoc.org/Download/PNS/Pain

**Figure 3. Summary of included trials**

Includes intervention studied and sample size

**Table 1. Summary of included trials**

Six trials were reviewed for safety and efficacy data.

**Figure 2. Visual Analog Scale**

Patients are asked to rate pain on a 10 cm line and the distance from no pain is considered the pain score.

**Figure 1. Wong-Baker FACES Pain Rating Scale**

Recommended for children three years and older. Child is asked to choose the face that best describes his/her own pain.