Review Article

Sickle Cell Disease Pain Management in Adolescents: A Literature Review

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Abstract:

Sickle cell disease (SCD) pain continues to emerge in adolescents. More than 98,000 individuals are believed to have SCD in the United States. In fact, 1 in 500 Black infants will be affected by SCD. Identifying standards of care for this unique population can improve pain management and treatment. A significant effect of vaso-occlusive crisis is a decrease in the quality of life in children. Therefore, pain management is multidimensional and includes pharmacologic, physical, and psychological strategies. A review of the literature was conducted to identify best practices regarding pain management in adolescents with sickle cell anemia. Key words such as *pain, pain management, adolescent sickle cell anemia*, and *acute sickle cell pain* were entered into databases to reveal qualitative and quantitative studies from 2009 to the present. Many of the research articles identified poor SCD pain management. Studies showed that acute SCD pain management is essential and should be evaluated and robustly managed to achieve optimum pain relief for patients. Acute SCD pain usually occurs as a result of vaso-occlusive crisis. Untreated acute SCD pain can result in morbidity and mortality in adolescents. Nursing knowledge is critical to reducing the stigma and improving management of SCD pain. Nurses play a vital role in the introduction of evidence-based practice within the clinical setting. In an effort to educate nurses and other health care professionals about SCD, this article is a literature review of studies concerning SCD and pain management in emergency rooms.

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BACKGROUND

Sickle cell disease (SCD) is characterized as the most prevalent genetic blood disorder affecting red blood cells worldwide (Myers & Eckes, 2012). SCD is believed to affect more than 98,000 individuals in the United States alone and 1 in 500 Black infants (Porter, Feinglass, Artz, Hafner, & Tanabe, 2012; Vijenthira et al.,

2012). SCD is characterized by recurrent, acute severe pain episodes due to vaso-occlusive crisis (VOC). The acute severe pain episode is caused by ischemic tissue resulting from blood vessel occlusion preventing blood flow (Vijenthira et al., 2012). VOC has been shown to decrease children's quality of life. Therefore, pain management is multidimensional and includes pharmacologic, physical, and psychological strategies. According to Vijenthira et al. (2012), treating VOC pain should occur early and be aggressive to prevent morbidity.

SCD patients seen in the emergency room (ER) wait at least 4 hours before receiving their first dose of pain medication (Haywood, Tanabe, Naik, Beach, & Lanzkron, 2013). SCD patients can have acute or chronic pain or a combination of both resulting in malformed sickle-shaped red blood cells. Sickle-shaped red blood cells cannot flow properly through the blood vessels and capillaries causing significant pain (Myers & Eckes, 2012). Both acute and chronic pain episodes require timely treatment.

SCD patients often are seen as drug seekers once they enter the ER (Haywood et al., 2013). Many of these patients manage their pain at home; however, an average of 197,000 ER visits occur each year, with a hospital admission rate of 29% (Porter et al., 2012). The cost for SCD patients seen in the ER for VOC has been estimated to be more than $356 million. Elkins, Johnson, and Fisher (2012) indicated that pain is a "significant and ubiquitous health problem, costing productivity, employment, income, reducing quality of life, degrading relationships, and negatively affecting physical and psychological well-being" (p. 294). Even with the guidelines and recommendations for pain management in patients experiencing VOC episodes there continues to be a gap with providing pain control among this group.

In part, 63% of nurses working with these patients believed addiction was a factor, and 30% were reluctant to administer a high dose of analgesic (Porter et al., 2012). Therefore, undertreatment of pain occurs in patients with SCD experiencing a VOC. Research by Meier and Miller (2012) suggests that although opioids were used for SCD patients with painful VOC episodes, drug dependency in SCD does not differ from other individuals in the general population without SCD. Dampier, Haywood, and Lantos (2011) indicated that drug seeking is often assumed due to the excessive use of opioids; however, it is somewhat uncommon among pediatric patients with SCD. In part, hyperalgesia from frequent opioid withdrawal or excessive use is more than likely the medical reason.

Often, SCD patients are not drug seekers but are seeking care due to the lack of "psychosocial support, poor coping skills, and inappropriate therapeutic expectations" (Dampier et al., 2011, p. 128). SCD pain management should be determined and will require multiple medications in which responses are made on a variety of neurochemical pathways. Wang, Kavanagh, Little, Holliman, and Sprinz (2011) found that 6% of patients with SCD die during childhood. Wang et al. (2011) suggested there is evidence between gaps and variations in quality of care that contributes to mortality in children affected with SCD. Also, poor outcomes are a factor in the quality of care in SCD.

PURPOSE

Nursing knowledge is critical to reducing the stigma and management of SCD pain. The purpose of this article is to investigate the best practices related to the management of SCD pain in adolescents. Identifying standards of care for this unique population can improve pain management and treatment. Nurses play a vital role in the introduction of evidence-based practice within the clinical setting. In part, nurses will serve as a catalyst providing scientific evidence to help them inform peers to facilitate change in adolescent SCD pain management.

SEARCH CRITERIA

A comprehensive literature review was performed using multiple databases including Medline, PubMed, Cochrane Database, EBSCOhost, CINAHL, and Health Source: Nursing Academic Edition. The original search yielded 27 appropriate articles. Key terms included *sickle cell disease, pain, adolescent, treatment, vaso-occlusive crisis, nursing,* and *emergency room*. Due to the limited number of retrieved studies related to SCD, other databases such as ProQuest, ProQuest Nursing and Allied Health Source were added to search areas of sickle cell pain in adolescents. This generated 4,134 studies and articles to collect the needed data for this literature review. Key terms were narrowed to include *sickle cell pain, nursing, pain, management, adolescents,* and *vaso-occlusive crisis*. These terms were entered into ProQuest and ProQuest Nursing and Allied Health databases. The results were narrowed by date of publication, to include articles published from 2009 to the present. The final nine studies selected included quantitative, qualitative, cross-sectional, randomized control trials, and comparative analysis dealing with SCD and pain management and were analyzed and selected based on sickle cell pain management.