# ARTICLE IN PRESS

Disease-a-Month ■ (2018) ■■■-■■■



Contents lists available at ScienceDirect

### Disease-a-Month

journal homepage: www.elsevier.com/locate/disamonth



# Oral health considerations for pediatric patients with sickle cell disease

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#### Introduction

Sickle cell disease (SCD) is a term given for a group of inherited red blood cell disorders that is caused by an abnormal hemoglobin, called hemoglobin S or sickle hemoglobin. It is the most prevalent genetic blood disorder worldwide. The two key characteristics are chronic hemolytic anemia and intermittent vaso-occlusion causing acute, severe pain episodes. SCD is a chronic disease that has detrimental effects on the entire body and requires a multidisciplinary team for management.

SCD usually does not present with symptoms until the first 3–6 months of life due to newborns still producing significant amounts of the protective fetal Hb (HbF), which inhibits the polymerization of HbS and protects against SCD.<sup>3</sup> However, splenic sequestration can begin as early as 3 months; thus penicillin prophylaxis should be started at 2 months until 5 years of age to help prevent pneumococcal infections. This is critical as children with SCD are at high risk for infections due to loss of splenic filtrate function, causing pneumococcal sepsis to be a leading cause of mortality among infants.<sup>4</sup> Consequently, high fevers (> 101–105°F) is a medical emergency in children with SCD because it can be the first clinical sign of bacteremia.<sup>3</sup> With the introduction of the pneumococcal conjugate vaccine (PCV), the rate of invasive pneumococcal disease (IPD) amongst children with SCD has dropped significantly.<sup>5</sup> Penicillin prophylaxis is usually discontinued after the age of 5 years due to no longer being a significant difference in the rate of IPD between the children who are and are not taking the prophylactic antibiotics at that age.<sup>6</sup>

Painful vaso-occlusive crises (VOC), also referred to as acute pain crisis, begin early in childhood as dactylitis and continues into adulthood as unpredictable painful episodes.<sup>4</sup> Most painful episodes are

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erratic, however it is thought that some triggering factors include infection, dehydration, extreme temperatures, hypoxia, physical or emotional stress and menstruation.<sup>4</sup>

Preventative care and meticulous oral hygiene is of utmost importance for patients with SCD for numerous reasons. Infection is the most common triggering event in a sickle cell crisis, thus any potential source of oral infection should be removed and oral infections must be treated aggressively. The research reveals controversial results on if prophylactic antibiotic therapy given at a young age is protective against dental caries. Research by Fukuda et al. state that the long-term penicillin prophylaxis in SCD patients delays the acquisition of mutans streptococci (MS), which results in a lower caries rate in primary teeth while taking the antibiotic therapy. Other studies reported no effect on the cariogenic microorganism levels; the inconsistent research suggests SCD patients require the same attention in regards to caries prevention as the general pediatric population.

#### **Dental management**

Prior to treating the pediatric patient with SCD in the dental chair, it is critical to complete a thorough medical history with questions concerning the severity of their SCD, medications (including the use of nonsteroidal anti-inflammatory drugs), disease related complications including, jaundice, splenectomy, bleeding disorders, renal issues, or growth impairment, the frequency of pain episodes and VOCs, their treatment for SCD, as well as consulting the patient's hematologist and pediatrician. Patients with severe SCD may have the option of being treated with a Hematopoietic Stem Cell Transplant (HSCT). Prior to receiving the HSCT it is important for the patient to have an oral exam and complete oral rehabilitation of any dental treatment needed.

#### Prevention

Infections are a leading factor in precipitating a sickling crisis; as a result, carious lesions that may lead to a dental infection should be treated aggressively. In the case of an acute dental infection, incision and drainage as well as intramuscular or intravenous antibiotics should be considered. If cellulitis develops a consult with the hematologist is warranted as the patient may require hospitalization.9 A rigorous oral health preventive plan should be implemented early for these patients 13 and can take place in both the dental and medical setting. Reinforcing the importance of oral health and meticulous oral hygiene by any treating physician (hematologist, pediatrician, etc) is paramount for a successful outcome for these children. Other preventative tools that treating physicians can utilize include dietary counseling and application of topical fluoride varnish. In the dental setting, along with the aforementioned, pit and fissure sealants and three-month dental recall visits should be considered for high risk patients.<sup>13</sup> SCD requires lifelong comprehensive care, making the education of parents, caregivers and the affected persons indispensable for improving quality of life and decreasing the morbidity and mortality of the disease. It is essential for the patient and family to understand the relationship between oral health and SCD and that poor oral health can lead to VOCs. The American Academy of Pediatric Dentistry (AAPD) recommends the establishment of a dental home care for all children by the age of 1 year. <sup>14</sup> A referral to a pediatric dentist from the hematologist or PCP by their first birthday can help reinforce the importance oral health and successful management of SCD.

#### Nitrous oxide

For routine dental care, appointments should not be scheduled during a crisis state and should be short to minimize stress. The use of epinephrine was once controversial as some authors believed that it may cause impairment of circulation through vascular occlusion. <sup>15</sup> However, there has not been any evidence to support the avoidance of epinephrine, thus the use of local anesthetic with 1:100,000 epinephrine in order to achieve hemostasis and profound anesthesia is considered safe

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