Oral health management considerations for patients with sickle cell disease

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A B S T R A C T

Sickle cell disease (SCD) can affect many systems in the body including the oral cavity. This necessitates modifications in oral health care management of these patients. The purpose of this paper is to make recommendations for management of SCD patients according to their oral health care needs based on the known findings.

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General management recommendations for patients with sickle cell disease in the dental practice

There is insufficient evidence in the literature to guide oral health practitioners on dental management of SCD patients.1 A systematic review revealed that there are no guidelines and insufficient information on how to treat dental complications and patients with SCD. The lack of guidelines can result in patients with SCD not receiving the appropriate treatment needed due to dental providers lack of knowledge on what complications to expect and how to manage these complications.
complications once they occur, and what treatment modifications are needed to accommodate these patients’ oral health needs. Neglecting oral health is common in patients with SCD for many reasons. Medical communities, as well as the patients, may fail to understand the correlation between oral health and their general health along with the impact poor oral health could have on their hematological disorder. Thus, there is a tendency to prioritize other more compelling health care needs over oral health. Low socioeconomic status and income disparities are factors that may delay patients from seeking dental care, along with dental care often being limited or excluded from health care insurance benefits. Consequently, if the patient neglects their oral health they become at a higher risk for caries and infections that could lead to more complicated dental treatment needs and higher chances of sickle cell crisis and emergency hospital admissions. It is essential that the treating provider is well informed and educated regarding the patient’s complete medical history including the stability of the disease, frequency of pain crises and SCD crises, SCD related complications, and any past and current medical treatment as well as all medications the patient is taking.

Prevention and early intervention

Prevention of complicated dental needs can be achieved by routine dental visits, and practicing good oral hygiene to prevent loss of teeth due to caries and infections. Thus, it is recommended that patients have periodic oral health screenings provided by a dentist at least every 6 months. Furthermore, the significance of oral health maintenance and healthy diet should be emphasized by all health care providers (primary care providers, hematologists, etc) at routine general health appointments and through oral health promotion programs in patients with SCD and in the community. A healthy diet, routine dental checkups and regular oral hygiene maintenance may help prevent or at least minimize oral health-related complications in patients with SCD. Other measures such as maintenance of proper hydration and avoidance of adverse climate conditions should be observed by the patient to decrease the chances of sickle cell crisis. Due to the increased RBC destruction and the RBCs shortened lifespan the patient’s physician may prescribe folic acid, Vitamin B12 or Vitamin B6 supplementation. Collaboration between the patient’s entire health care team, including the hematologist and dentist is important to ensure the patient is receiving the proper treatment.

Stress reduction and pain management strategies in dental practice

Stress is a well-known factor that provokes a sickle cell crisis, thus minimizing stress during dental treatment is crucial. Short morning visits are recommended for SCD patients. An anxiety assessment can be performed at the initial visit by asking questions on the patient’s anxiety and feelings in regards to the anesthesia and procedure, as well as on past dental experiences in order to help accurately assess the patient’s level of anxiety prior to treatment so proper anxiety-reducing treatments can be used when needed. If the patient’s SCD is under therapeutic control, outpatient procedures can be performed without complications during non-crisis periods by using topical anesthesia followed by local anesthetic such as 2% lidocaine with 1:100,000 epinephrine. During a sickle cell crisis only acute infections or problems such as trauma should be treated and definitive treatment should be postponed until the patient is in a non-crisis state. For mildly anxious patients, prescription of anxiolytics and sedatives can be used to help manage patients’ anxiety with midazolam or valium. However, barbiturates and narcotics should be avoided as these drugs may cause respiratory suppression leading to hypoxia, acidosis and ultimately a sickle cell crisis. It is imperative to consult with the hematologist prior to any sedation procedure. For highly anxious patients or patients with extensive multiple dental or surgical procedures general anesthesia may be recommended. In those scenarios, patients are referred to a hospital with a hematology consult service and may be admitted prior to the procedure. It is recommended that a preoperative level of hemoglobin be evaluated, and a blood transfusion may be warranted to decrease the HgbS to < 30%, which will be determined by the patient’s hematologist.
Care should be taken to keep the patients well hydrated and oxygenated, as well as maintain normal temperature and to avoid acidosis and infections before, during, and after the surgical procedure. For postoperative pain management acetaminophen or acetaminophen with codeine is recommended. Although non-steroidal anti-inflammatory drugs (NSAIDs) have the potential to be an important component of the pain management for SCD patients, NSAIDs should be used cautiously, especially for those patients with renal, gastrointestinal and cardiovascular risks. Aspirin is not recommended due to its possible adverse effects including decreased platelet function resulting in increased bleeding, acidosis and bone marrow suppression. Anti-inflammatory analgesics should be used at the lowest effective dose and for the shortest duration in this patient population and regular monitoring of potential side effects is required for all patients receiving long-term treatment. SCD patients are often perceived by health care practitioners as “drug seekers”, which result in delaying effective pain relief and often resulting in under-treatment that can prolong suffering and result in repeat emergency visits. Health care professionals, including dentists, should remember that pain is a feature of sickle cell crisis and should be compassionate about the patients’ complaints.

Infection management

Patients with SCD are at a higher risk for infections, including dental infections, than the general population. The increased risk of infection in patients in SCD is due to multiple factors including autosplenectomy at a young age as well as during a crisis where the macrophages are unable to function normally against the infectious agent due to the numerous sickled RBCs. Additionally, the neutrophils do not function properly in terms of adherence, migration and bactERICidal function. Dental infections may be responsible for precipitating a sickle cell crisis, additionally they may exacerbate an already exiting crisis through inflammatory mediators which may lead to visits to the ED and admission to the hospital. Prevention and early management by removal of any potential source of infection from gingival, periodontal or endodontic origin are essential. If dental infections should occur, they must be treated aggressively with both local and systemic measures. Antibiotic prophylaxis is recommended for invasive dental and major oral surgical procedures. Prophylactic antibiotics may prevent the precipitation of vasculo-occlusion crises, osteomyelitis and systemic infections originated by dental infections. Hydroxyurea is a common medication used to treat SCD and can trigger neutropenia and thrombocytopenia; which may be another risk factor for infection, patients taking this medication should have a complete blood count taken before any invasive dental procedure.

Restorative and prosthodontic management

Due to tooth hypomineralization and diminished oral hygiene, SCD patients are identified as extreme high-caries risk patients based on caries management by risk assessment (CAMBRA). The higher the severity of the risk factors, the greater the intensity of protective factors that must be implemented to reverse the caries process. These protective factors include a variety of products and interventions that will promote teeth remineralization and protection of the patient's dental and oral health. These protective factors include sealants, antimicrobials (chlorhexidine) or antibacterial (including xylitol), regular use of fluoride containing products such as toothpastes, oral rinses and the topical application of fluoride varnish and calcium phosphate agents. Thermoformed appliances can be used for the application of fluoride containing gel or paste to reduce the hypersensitivity and prevent caries, further, to facilitate the oral hygiene management. Periodic usage of chlorhexidine, one week per month, can be helpful to prevent oral and dental infection. Regarding replacing missing teeth with dental implants, so far, no literature is available to support surgical implant therapy in SCD patients. A very limited number of studies have evaluated the effect of other bone diseases on dental implant outcomes. Understanding the pathological
events in bone of SCD patients, it is prudent to say that implant therapy is not recommended for these patients. The hemoglobin disorders, medullary hyperplasia and abnormal trabecular spacing may negatively affect bone-to-implant contact (BIC). In addition, reduction of bone mechanical properties and delayed bone healing may impair the osseointegration process and long term success of implant therapy.29 Susceptibility for infections, decreased blood supply due to vaso-occlusion events in the jaw bones, boney infarcts makes implant therapy not a suitable treatment modality. However due to lack of evidence in literature we cannot speculate that is it an absolute contraindication. It is in the best benefit of the patient is to explore less invasive prosthetic options and provide conservative treatment with the least surgical trauma involved. Removable partial dentures are a common conservative, less expensive prosthetic treatment option for partially edentulous patients. Fixed partial prosthesis (crown and bridge work) may have unpredictable prognosis since SCD patients are at risk for various perioperative complications, and pulp necrosis of unknown origin.30 Susceptibility to caries remains a challenge in long-term success of this treatment modality. However, in a well-maintained patient, this can be a good option. Fig. 1 represents a case of an African American female patient who received early intervention and is under regular dental maintenance care. This is an example of a reasonable, minimally invasive and successful prosthetic treatment.

Orthodontic management

SCD appears to be a risk factor for moderate and potentially severe dental malocclusion.32 Pathological changes in bone structure and morphology due to characteristic compensatory bone marrow expansion can be expected and observed radiographically.32 Children with SCD often have delayed tooth eruption and enamel hypoplasia of teeth. The most common craniofacial bone abnormalities in SCD individuals as reported in the literature were maxillary protrusion, overjet, overbite, retrusion of mandible and large trabecular bone. They may also have frontal bossing, which may compromise the patients’ appearance and self-confidence.33 Referral to an orthodontist is required for treatment of occlusal and skeletal discrepancies. The orthodontic treatment will enhance facial and dental features by reducing the overjet and overbite to normal values and the relieving crowding, which makes practicing oral hygiene easier for the patient. Antibiotic prophylaxis is recommended prior to any procedures that may result in bleeding and promote bacteremia. Orthodontic procedures that induce bacteremia should be avoided which may happen when placing and removing separators or orthodontic mini-plates.34 Extraction of asymptomatic teeth is contraindicated in patients with SCD and should be considered only when these teeth are unrestorable. Extra-oral appliances should be considered when correcting class II malocclusion in patients with SCD in lieu of extractions, use of miniplates, or orthognathic surgeries.32,33,35 During orthodontic treatment, the bone response, pulpal health, and the systemic condition should be monitored. Similar to other dental procedures, emotional stress should be avoided and care to have adequate levels of oxygenation, good hydration and ambient temperature should be provided.

Fig. 1. Post dental treatment replacing missing teeth in a Sickle Cell Disease African-American female patient.
In general, clinical appointments during chronic stages of the disease, light orthodontic forces, frequent visits and potentially longer overall treatment time are needed for SCD patients.\textsuperscript{36} In addition, long extensive procedures should be avoided and appointments is preferably scheduled in the early morning.\textsuperscript{37} The oral hygiene status of the patient should be put under strict control to avoid any episodes of gingivitis during the treatment.

**Conclusion**

This paper presented special recommendations when providing oral health care for sickle cell patients. Some dental aspects were well addressed in the literature while others are lacking evidence such as implant therapy. Studies are needed to provide evidence regarding implant therapy in SCD patients. Early intervention, prevention, maintenance of oral health are essential in stable and long-term success of dental treatment. Patient hematological status and modification of dental therapeutic modalities may be necessary for these patients using clinical judgement and available evidence. Close collaboration between dentists and hematologists and sickle cell clinics are essential in overall management of SCD patients.

**References**


