

## Sickle Cell Disease's Orofacial Manifestation and Dental Management: A Scoping Review

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### Abstract

A growing global health issue, sickle cell disease (SCD) has seen tremendous advancements in treatment, particularly after 2017. The dental treatment for sickle cell disease was not, however, included in any systematic evaluations of clinical trials (SCD). This article seeks to describe the oral characteristics of sickle disease and talk about oral management techniques that dental professionals can use as a guide. With the help of Web of Science, Google Scholar, and PubMed, a thorough literature review was carried out. The tactics for searching were created to include publications from January 2010 to March 2020. Keywords were used to identify numerous abstracts. These abstracts were subsequently examined, revealing details on the oral health characteristics of SCD manifestation.

A narrative review that enumerates every facet of the oral manifestation in persons with SCD was created based on all of these articles and clinical practise. According to the study's findings, there is clear evidence pointing to a developmental enamel deficiency that causes hypoplasia and increases a person's susceptibility to dental caries. Another significant finding of this review was that individuals with SCD experience a vaso-occlusive crisis in the dental pulp's microcirculation, which can cause both symptomatic and asymptomatic pulpal necrosis without odontogenic disease in an otherwise healthy tooth.

The study also discovered that employing a multidisciplinary approach has a significant role in managing persons with SCD and that early detection, intervention, and prevention are vital for improved oral health care. Chronic general health issues plague sickle cell disease patients. Their primary attention shifts to the haematological condition, making poor oral health secondary and, at most, raising the risk of dental caries. This study offers guidelines for better dental therapy of SCD patients as well as a general description of the oral manifestations of SCD. Dental professionals frequently misinterpret SCD patients, and they are unable to provide effective care because they lack the necessary information and criteria. In order to provide better dental care, this paper tries to emphasise the necessary steps.

**Keywords:** Sickle cell; Systemic health; Evidence-based dentistry; Caries risk; Malocclusion pulp necrosis; ischemia; Osteomyelitis; mental nerve neuropathy; Oral pain

### Introduction

Red blood cells that are affected by Sickle Cell Disease (SCD) have abnormal shapes and degrade rapidly. The haemoglobin, which delivers oxygen to every cell in the body, is aberrant in SCD. Haemoglobin S, the aberrant haemoglobin, has a poorer functional capability and contributes to a number of systemic problems. The condition gets its name from haemoglobin S, which causes red blood cells to take on a sickle- or crescent-shaped form. One of the most widespread inherited blood disorders in the US is SCD. Over 100,000 Americans are thought to have SCD, according to the Centers for Disease Control and Prevention (CDC) [1]. SCD affects an unknown number of people, but it has been estimated that 1 in every 365 African American babies is born with the condition, and 1 in every 13 African Americans also possesses the sickle cell trait [2]. Based on data from 13 states, the estimated incidence of sickle cell trait in the United States was 73.1 cases per 1,000 black infants, 3.1 cases per 1,000 white infants, and 2.2 cases per 1,000 infants of Asian or Pacific Islander descent. Within 13 states, the incidence estimate for Hispanic ethnicity was 6.9 cases per 1,000 Hispanic new-borns. The World Health Organization (WHO) estimates that 5% of the world's population carries the trait genes for haemoglobinopathies, with sickle cell disease and thalassemia being the two most common. Between 2010 and 2050, the number of infants born with sickle cell disease is projected to increase by almost 30%. In lower- and middle-income nations, SCD affects a large proportion of new-borns. Because of delayed diagnosis and treatment, most affected children pass away during the first few years of life, with excess mortality rates as high as 92% [3].

Normal adult haemoglobin (Hb-A) molecules have four polypeptide chains, two of which are alpha units and two of which are beta units. One heme group, which serves as the oxygen molecule's binding site, is present in each chain. Various amino acid sequences in both chains fold to produce different three-dimensional structures. The interactions between the four chains are monovalent. In the haemoglobin beta chain, a point mutation in SCD converts glutamic acid to valine [4]. Red blood cells that have this form of aberrant haemoglobin, also known as Hb-S, become rigid and have an irregular shape. The red blood cell is deformed into a sickle or crescent shape rather than its typical round disc shape.

Red blood cells typically live for 90–120 days, however in SCD; they only last for about 10 days [5]. Red blood cells that have sickle-shaped haemoglobin breakdown early in the spleen, resulting in fewer total red blood cells, sickle cell anaemia, and hyperbilirubinemia. Fatigue, irritability, dizziness, light-headedness, tachycardia, and shortness of breath are just a few of the symptoms of an oxygen deficit that are brought on by sickle cell anaemia. The main molecule that transports

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oxygen to all of the cells throughout the body is haemoglobin, which is found in red blood cells (RBCs). Jaundice, commonly known as a yellowing of the eyes and skin, is another side effect of the fast breakdown of RBCs known as haemolytic anaemia [6].

## Materials and Methods

The literature on sickle cell illness and its oral symptoms was evaluated for this investigation. Controlled clinical investigations, retrospective studies, experimental research, and reviews are all included in the literature that discusses SCD, oral manifestations, and dental care [7]. The subject "What the diverse oral manifestation seen in the patients with the SCD and how these patients are can be better managed" was the focus of a narrative review that was built reporting items. Patients with SCD make up the population (P); common dental operations performed in a clinical context serve as the intervention (I); healthy controls serve as the control group (C); and dental management strategies serve as the outcome measure (O). The purpose of this article is to describe the oral characteristics of SCD and to go over oral management techniques that dental professionals can use as a reference [8].

## Discussion

Oral injuries caused by oneself can be deliberate, unintentional, or the result of an odd habit. These wounds are frequently caused by a patient's fingernail or a foreign item that frequently harms the gingival tissue or teeth. The severity of self-injurious conduct varies, ranging from mild fingernail biting to major self-mutilation. To the best of the authors' knowledge, no one has yet documented the current case in which the usage of a metallic compass resulted in mechanical injuries. These cases offer yet another chance to underline the importance of a thorough history that uncovers the more nuanced facts regarding aetiology. A sound treatment strategy might result from a thorough case history and accurate radiography interpretation [9].

The pulp chamber and the root canals were said to include a variety of foreign things, including beads, stapler pins, metal screws, darning needles, and pencil leads. In the root canals of anterior teeth left open for drainage, Grossman and Heaton reported finding indelible ink pencil tips, brads, a toothpick, adsorbent points, and even a tomato seed. A 12-year-old Japanese boy has reported finding a plastic chopstick lodged in an interrupted supernumerary teeth in the premaxilla [10].

A radiograph may be useful for diagnosis, particularly if the foreign body is radiopaque. Hunter and Taljanovic included the parallax views, vertex occlusal views, triangulation techniques, stereo radiography, and tomography as the many radiographic approaches to be used to locate a radiopaque foreign object. Different materials' ability to attenuate X-rays determines how visible they are on plain radiographs; foreign bodies may be seen based on their inherent radio density and closeness to the tissue in which they are lodged. Radiographic images of metallic items, including most animal bones and all foreign bodies composed of glass, are opaque unless they are made of aluminium.

Clinical behaviour in patients with penetrating wounds depends on the type of foreign substance; inert materials like steel and glass might

not generate enough inflammation to require removal. However, it is imperative to remove organic foreign materials since they frequently cause subsequent infection, the formation of an abscess, and a fistula. The different foreign bodies in our case included the tip of a metallic compass, a stapler pin, an air conditioner copper chip, and needle fragments that needed to be promptly removed so that the right treatment could be administered [11].

## Conclusion

This article offers suggestions for improved care and an understanding of the underlying aetiology of such issues as well as a general description of the oral manifestations of SCD. Dental professionals frequently misinterpret SCD patients, and they are unable to provide effective care because they lack the necessary information and criteria. In order to provide better dental care, this paper tries to emphasise the necessary steps. It is crucial that a team effort is made with the assistance of a haematologist, a dentist, and a primary care physician. For SCD patients, better oral health care requires early detection, intervention, and prevention. To show which therapy approaches are most successful, more study is encouraged.

## Conflicts of Interest

None

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