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Original article

Dental tissues of sickle cell anemia and its impact on the quality of life related to oral health

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ABSTRACT

Objective: The aim of this study was to determine the association between dental tissues and sickle cell anemia (SCA) and how it impacts the quality of life related to oral health.

Materials and Methods: It was a cohort study of 154 Congolese participants with and without SCA conducted in the dental service of SCA at the Yolo Center, Kinshasa, aged at least 6 years and without a history of clinically severe conditions (hospitalization and blood transfusion), who were regularly monitored. The inclusion criteria were the diagnosis confirmation of SCA at the health service in a period of at least 6 months before enrollment in this study. Dental tissues were assessed by a clinical examination using a dental mirror and probe. The index of Decayed-Missing-Filled Teeth (DMFT) was used to assess the dental state of the participants. For Oral Health-related Quality of Life (OHRQoL), the Congolese versions of the perception questionnaires, modified from the Oral Health Impacts Profile (OHIP-23), were used for participants. Each question had to be answered by yes or no, depending on whether the participant was satisfied (outcome = 1) or dissatisfied (outcome = 0) about an oral health-related quality of life.

Results: Of the 154 participants, aged from 6 to 64 years, with a mean age of 19.5 ± 7 (SD) years, 96 presented with SCA and only 68 were correctly followed; 102 did not present SCA and only 86 were correctly followed. The DMFT and dmft indexes were higher in the SCA group, being 2.9 and 2.5, respectively. The difference between the SCA group and the

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control group was significant for decayed teeth, missing teeth, filled teeth and no caries. Of the different dimensions of quality of life that were compared between the SCA group and control group, 15 of 23 items were statistically significant.

Conclusion: The present study strongly confirmed an association between dental caries and missing teeth with sickle cell anemia. Secondly, the quality of life for SCA participants seems to be poor, compared to the control group.

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Introduction

Sickle Cell Anemia (SCA) is a qualitative hemoglobinopathy characterized by the substitution of glutamic acid for valine at the sixth codon of beta chain s globin.¹ Global estimates suggest that more than 312,000 infants are born with SCA each year.² The United States and Europe represent 2 % of annual SCA births worldwide.³ The vast majority of sickle cell births occur in developing countries, with an estimated number of 230,000 annual SCA births in sub-Saharan Africa. The reported mortality reaches up to 92 %.^{4,5} The Democratic Republic of the Congo (DRC) has the highest incidence of SCA patients in Africa after Nigeria.^{6–8} The number of children born with SCA is expected to grow by nearly 30 % from 2010 to 2050.⁷

In SCA, the life span of red blood cells is severely diminished from the usual 90 to 120 days to about 10 days.⁹ Due to atypical hemoglobin and their sickle shape, red blood cells break down prematurely in the spleen, causing fever and hyperbilirubinemia. This leads to a structural modification responsible for changes in the stability, solubility and structure of erythrocytes, which take on a sickle shape when exposed to low oxygen tension, acidosis or dehydration.¹⁰ Hemoglobin in the red blood cell is the main molecule that delivers oxygen to all the cells throughout the body. SCA results in multiple symptoms of oxygen deficit, including fatigue, irritability, dizziness, lightheadedness, tachycardia and shortness of breath.

In the literature, SCA has been linked to dental caries, tooth erosion, hypomineralization, hypercementosis, pulp stones, and asymptomatic pulp necrosis.¹¹ Mutombo *et al.*; found that the most common oral manifestations of SCA are mucosal pallor, yellow tissue coloration, disorders of enamel and dentin mineralization, changes to the superficial cells of the tongue; multiple caries and periodontal disease that can lead to an odontogenic abscess.¹² Additionally, frequent admission drug treatments and diminished oral hygiene are high-risk factors for caries in SCA patients.¹³

Oral health is part of general health and recognized as an essential component of quality of life (QoL) that may affect the general health.¹⁴ The quality of life and the familial conditions are critical in determining the process of health and disease.¹⁵ Several conditions affect the quality of life¹⁶ related to oral health in the daily lives of many patients. Many of them throughout the world, especially the poorest or those with inadequate oral hygiene are still affected by oral problems, such as caries and periodontal diseases.¹⁷

The aim of this study was to determine the effect of SCA on dental tissues and oral health-related quality of

life of the patients treated at the dental service of the SCA Center at the Yolo Center, in relation to the control group.

Materials and methods

Study design and sampling

A cohort random study with a non-probabilistic sampling design of Congolese participants with and without Sickle Cell Anemia (SCA) was conducted at the dental service of SCA at the Yolo Center Kinshasa. The SCA dental service at the Yolo Center is the only institution accredited by the Congolese health ministry of the government of the Democratic Republic of the Congo. The participants' medical records were obtained from the interview, dental examination and biological examination or blood examination report performed before or after treatment.

A total of 214 participants from January 2020 to December 2020 were invited to participate in the study. Of these, 154 agreed to participate (adhesion rate = 72 %). Only those participants who had completed all items on the clinical questionnaire and measuring instruments (OHIP-C, DMFT), aged at least six years, were included in the final sample. They were randomly assigned to 2 groups, an SCA group and a control group of healthy participants.

The inclusion criteria for the SCA patients were as follows: the diagnosis confirmation of sickle cell anemia for a period of at least six months prior to enrollment, hemoglobin electrophoresis revealing a genotype HbSS, the determination of the hemoglobin concentration analysis¹⁸ and the absence of painful crisis at the time of the survey, of medical conditions other than SCA, of emergency dental appointments in the past six months, or of a history of clinically health severe conditions (hospitalization and blood transfusion), being regularly monitored by the dental service of SCA at the Yolo Center.

The eligibility criteria for the healthy participant (control group) included the absence of organic, physiologic, or psychiatric disturbances, of intellectual disability, or of emergency dental appointments in the past six months. The exclusion criteria were the inability to meet the above requirements and refusal to participate in the study. The patients enrolled in the groups (SCA group and control group) and those lost to follow-up and their reason is detailed in the flowchart of patients enrolled and randomly followed in the study (Figure 1).

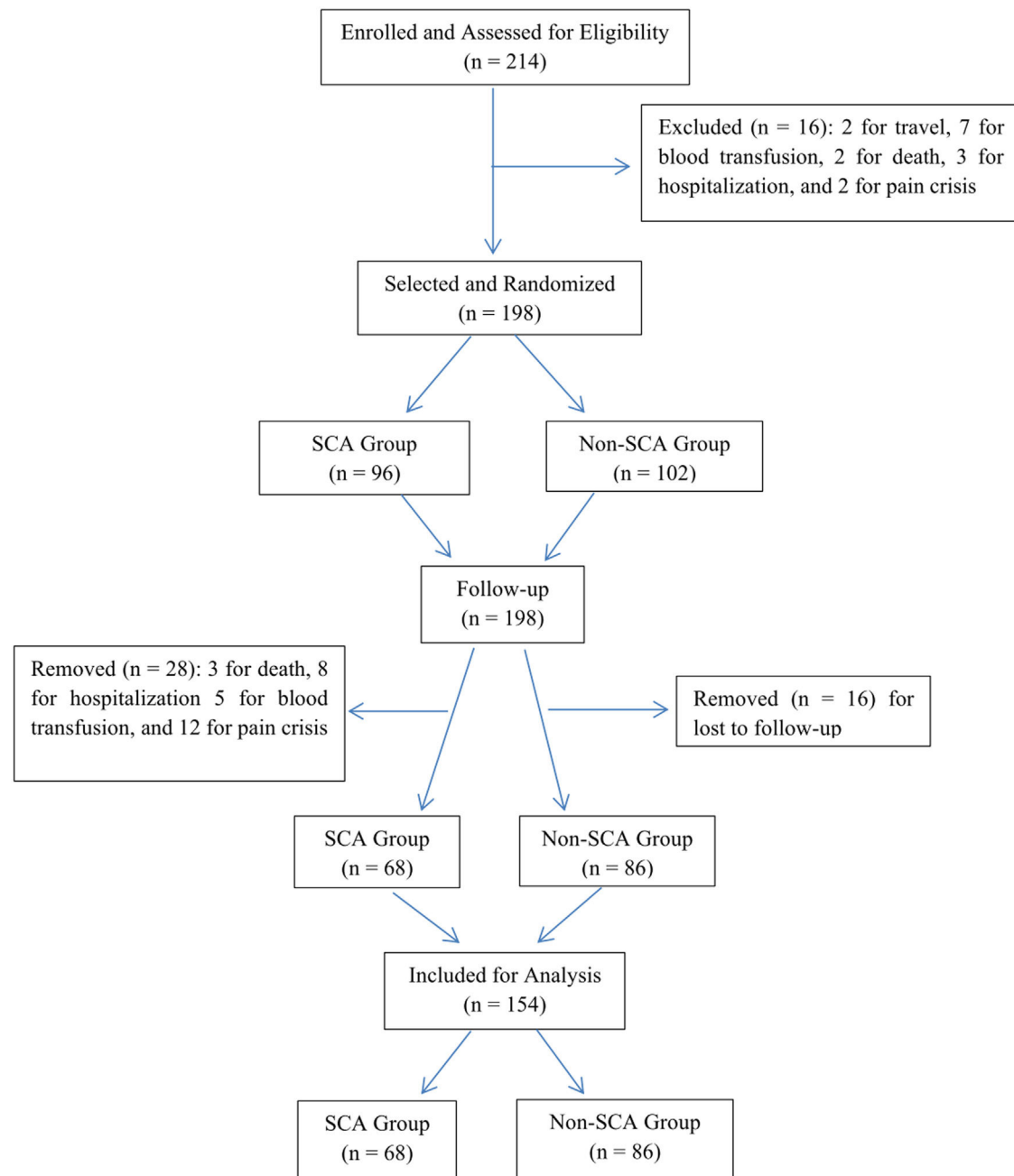


Figure 1 – Flow chart of patients enrolled and randomly followed in the Study.

Study variables

The study variables were gender, age, visiting dental service to predict economic situation, dental status (decayed teeth, missing teeth and filled teeth), functional limitations, psychological discomfort, physical inability and physical pain were evaluated.

Data collection and measurements

Interviews and dental exams were performed in the dental service room during the day. Data on the demographics and socioeconomic characteristics were collected through direct

questioning of the participants themselves or their parents or guardians. An intra-oral examination on each patient using a disposable mirror, CPI probes and gauzes was performed by 2 experienced and qualified periodontologists.

The assessment of the dental status was made by a clinical examination using a dental mirror and probe. The index of DMFT (Decayed-Missing-Filled Teeth), described by Klein et al.,¹⁹ was used to assess the dental state of the participants.²⁰ Score 0: lack of caries, Score 1: Presence of caries, Score 2: Caries filled, Score 3: Filling without caries, Score 4: Loss of teeth before prosthodontics restauration, Score 5: Loss of teeth after prosthodontic restauration and Score 6: Loss of tooth due to the periodontal diseases after prosthodontic restoration.

During the clinical examination, the dentist wore personal protective equipment (PPE) that met biosecurity standards.

As for the Oral Health-related Quality of Life (OHRQoL), the Congolese versions of the perception questionnaires,²¹ modified from the OHIP-23²² were used for participants. The questionnaires were prepared in the French language and helped to conduct a face-to-face pilot interview with 15 patients with Sickle Cell Anemia. After a normality pretest, data were analyzed with the Kolmogorov–Smirnov test and Cronbach's α correlation coefficient, with $p \geq 0.05$, indicating the survey had normal data distribution. The results of the pilot study were not included in the present study. Each question had to be answered by yes or no, depending on whether the participant was satisfied (outcome = 1) or dissatisfied (outcome = 0) about a specific aspect of his or her oral health-related quality of life. The overall oral health-related quality of life per participant was represented by the sum of the outcomes of the individual questions. The questions are similar for both groups.

Statistical analysis

Statistical analysis was performed using the Statistical Package for the Social Sciences (SPSS version 23.0). Descriptive and univariate analyses were performed separately for the SCA group and control group (healthy participants). The responses to categorical questions by the group were compared using either a chi-square test. The ANOVA test was used to compare medians of continuous variables between the SCA and control groups. The $p < 0.05$ was considered a level of statistically significant difference.

Ethical aspects

This study was authorized by the Research Ethics Committee of the National Center of Research for Dental Sciences in DR Congo-CNRSBD, which approved this study under the protocol number CNRSBD1504.215 and conducted according to the tenets outlined in the Declaration of Helsinki and to the

STROBE guidelines. The study included only individuals at least 6 years of age who agreed and signed the Free and Informed Consent Form. Authorizations for the use of the instruments were acquired together with the authors (OHIP-23) and the competent guardian.

Results

Of the 214 participants who were included, only 154 remained after correct follow-up until January 2021 (Figure 1) and constituted the final sample, from which the data were collected and statistically analyzed.

There was a male predominance in the SCA group and a female predominance in the control group. The most represented age group was ≤ 10 years in the SCA group and the age group of 11 to 20 years in the control group. The most frequent education level was primary school in the SCA group and middle school in the control group. A higher income was found in the control group. The difference between the SCA group and the control group was significant, as described in Table 1. The DMTF and dmft indexes were higher in the SCA group. The difference between the SCA group and the control group was significant for all the dental status, as described in Table 2. Among the different dimensions of quality of life that were compared between the SCA group and the control group, 15 out of 23 items were significant, as described in Table 3.

Discussion

Sickle Cell Anemia (SCA) has several common oral manifestations. In the Democratic Republic of the Congo, out of the studies investigated by,^{1,4,8,12} not one evaluated the SCA for dental tissues and oral health-related quality of life.

Table 1 – Socio-demographic characteristics of the sample.

Above 'lost' should be changed to « loss ».

Characteristics		SCA Group n (%)	Control Group n (%)	P-Value
Gender	Male	37 (54)	29 (34)	0.244
	Female	31 (46)	57 (66)	
Age-groups (years)	≤ 10	25 (37)	18 (21)	0.005
	11 - 20	22 (32)	21 (24)	
	21 - 30	13 (19)	11 (13)	
	31 - 40	6 (09)	13 (15)	
	40 - 50	2 (03)	12 (14)	
	51 - 60	0 (00)	5 (06)	
Education level	61 - 70	0 (00)	6 (07)	0.534
	illiterate	0 (00)	0 (00)	
	Primary School	33 (49)	29 (34)	
	Middle School	18 (26)	36 (42)	
	High School	7 (10)	8 (9)	
Family Income in Dental visit	University	10 (15)	13 (15)	0.002
	US\$ (SD)/month	161 \pm 71	253 \pm 86	
	Yes	15 (22)	50 (58)	
	No	53 (78)	36 (42)	0.022

SCA: Sickle-Cell Anemia, US\$: United States Dollars.

Table 2 – Dental status of the sample.

Dental status	SCA Group n (%)	Control Group n (%)	P-Value
Decayed teeth	664 (31)	334 (12)	0.000
Missing teeth	807 (38)	253 (09)	0.000
Filled teeth	541 (21)	518 (19)	0.041
No caries	221 (10)	1680 (60)	0.000
Total	2143 (100)	2785 (100)	
DMFT index	2.9	1.2	
dmft index	2.5	1.7	

SCA: Sick Cell Anemia; DMFT index: Decay-Missing-Filled Teeth index for permanent teeth; dmft index: decay-missing-filled index for temporary teeth.

The result showed a significant association between dental caries and missing teeth and a more negative impact on the QoL in patients with SCA, as was similar to other authors,¹⁸ meaning that, although most oral problems do not represent an immediate risk of death, they are responsible for decreasing the QoL of individuals, as they prolong states of pain and suffering and cause functional, esthetic, nutritional and psychological problems. Additionally, the morbidity for pain crises is a risk factor for caries and missing teeth.^{4,23} The minor occurrence of dental caries and missing teeth in the control group may be justified by their high frequency of attending dental visits, while those of the SCA group do not attend dental visits. These participants received major healthcare surveillance provided via access to dental

treatment at the SCA Center. This shows the great resilience capacity of patients with chronic diseases, including SCA, to maintain good oral health and to attend periodic annual dental visits.^{24,25}

Certain authors reported that the greater incidence of dental caries in patients with SCA is associated with a poorly structured family context, as well as worse health conditions in socially marginalized areas.¹⁵ A higher income was found in the control group (US\$ 253 ± 86). The difference between both groups was significant for family income and dental visits. The age of the patients with dental caries and missing teeth in SCA maybe play an important role. The behavior of older patients may differ from that of the younger ones. Teenagers can see the supervision of their parents as a threat to their growing desire for independence, resulting in resistance to the appropriate behaviors for health.²⁶ The knowledge of the association between dental caries and tooth loss in the SCA group allows dentists to gain a greater understanding of the problem and the role of dentists in overall health. Regardless of the effect of SCA on dental tissues, the oral health of this population needs a preventive measure against infections that could precipitate a vaso-occlusive crisis with other complications.^{12,27} However,²⁸ found that SCA does not predispose an individual to dental caries. Prevention of disease, disability and suffering should be a primary goal of each society that hopes to provide a decent quality of life for its people. Prevention on the community- or population-based level is the most cost-effective approach and has the greatest impact on a community or population, whether it is a school, neighborhood, or nation. Many different approaches to preventing

Table 3 – Oral health impact profile of sample.

OHIP-23 Items		SCA Group		Control Group		P-Value
		No n (%)	Yes n (%)	No n (%)	Yes n (%)	
Functional limitations	Mastication	26 (32)	42 (68)	53 (62)	33 (38)	0.004
	Bad pronunciation	24 (35)	44 (65)	77 (90)	23 (10)	0.003
	Affected appearance	31 (46)	37 (54)	86 (100)	0 (0)	0.003
	Breathing	23 (34)	45 (66)	81 (94)	5 (6)	0.000
	Bad taste	39 (57)	29 (43)	79 (92)	7 (8)	0.003
	Affected food intake	46 (68)	22 (32)	83 (97)	3 (3)	0.002
	Difficult adaptation	41 (60)	27 (40)	80 (93)	6 (7)	0.002
	Pain in the jaw	17 (25)	51 (75)	76 (88)	10 (12)	0.001
Oral symptoms	Dental sensitivity	26 (32)	42 (68)	80 (93)	6 (7)	0.001
	Dental pain	24 (35)	44 (65)	69 (80)	17 (20)	0.004
	Pain during the meal	25 (37)	43 (63)	77 (90)	9 (10)	0.423
Psychological discomfort	Anxiety/unrest	30 (44)	38 (56)	64 (74)	22 (26)	0.142
	Disquiet/Miserable	20 (29)	48 (71)	79 (92)	7 (8)	0.002
	Esthetic	43 (54)	31 (46)	82 (95)	4 (5)	0.005
Physical Inability	Unclear speech	43 (63)	57 (37)	71 (83)	15 (17)	0.003
	Diminished taste	27 (40)	41 (60)	67 (78)	19 (22)	0.322
	Dental brushing	29 (43)	39 (57)	60 (70)	26 (30)	0.304
	Food avoidance	36 (53)	32 (47)	73 (85)	13 (15)	0.286
	Poor diet	33 (49)	35 (51)	83 (97)	3 (3)	0.318
Physical pain	Eating difficulty	32 (47)	36 (53)	77 (90)	9 (10)	0.324
	Avoid to laugh	23 (34)	45 (66)	67 (78)	19 (22)	0.246
	Lack of appetite	21 (31)	47 (69)	78 (91)	9 (8)	0.001
	Upset stomach	10 (15)	58 (85)	86 (100)	0 (0)	0.001

OHIP: Oral Health Impact Profile; SCA: Sick Cell Anemia.

dental diseases exist and the most cost-effective method is health education.

The quality of life for patients with SCA is critical in determining the process of health and disease. There was a difference in the OHRqOL among the SCA and control groups, similar to that of Ralstrom et al.²⁹ This study only included participants who were not suffering from a painful crisis at the time of the survey, medical conditions other than SCA and emergency dental appointments in the past six months. These facts may have led to evaluating only participants in good health, masking the OHRqOL during the acute pain that is a characteristic of SCA. The concept of quality of life related to oral health requires consideration not only of factors, such as malaise, pain, or functional changes, but should also include emotional aspects and social functions associated with oral health. Oral health is a significant component in the general quality of life of people, with important implications for their health status. The SCA severity is associated with the vasculature vaso-occlusion and may account for the pain experienced.^{30,31}

Our findings related to the OHIP-23 showed that the participants enrolled in the control group were most satisfied with the oral health-related quality of life than the participants of the SCA group. The difference between both groups was significant for 15 out of 23 items: mastication, bad pronunciation, affected appearance, breathing, bad taste, affected food intake, difficult adaptation, pain in the jaw, dental sensitivity, dental pain, disquiet/miserable or unhappy, esthetic, unclear speech, lack of appetite and upset stomach. Lower income may be associated with lower oral health, as well as lower access to preventive health care information.^{32,33}

The main limitation of this study was the fact we examined a specific population from Kinshasa, the capital of the Democratic Republic of the Congo. In addition, it is important to mention that the present results are of insufficient conclusiveness because only one oral index for clinical evaluation (DMFT) was used, as well as the fact that the analyses of some different domains related to quality of life were not included, but will be necessary for future studies. Prospective studies with larger samples may provide a better understanding of the relationship between SCA and dental tissues and the oral health-related quality of life.

Conclusion

Considering the findings of our study, dental caries are associated with sickle cell anemia and they negatively impact the quality of life. To prevent the occurrence of dental caries and tooth loss and to avoid complications in the general health of patients with SCA, the inclusion of dental visits and dental care in the management strategy of patients with SCA should be considered. The negative impacts that poor oral health of the population have on their QoL make this issue a relevant problem for public health. Consequently, dentists have an important role in preventing health complications in patients with SCA.

Conflicts of interest

The authors declare that there is no conflict of interest regarding the publication of this article.

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