



## Multidisciplinary management of homozygous sickle cell patients: Dental treatment pathologies and needs

 Yolande Koffi-Gnagne,<sup>1</sup>  Stéphane Xavier Djolé,<sup>1</sup>  Marie Chantal Avoaka-Boni,<sup>1</sup>  
 Guillaume Loukou,<sup>1</sup>  Douni Sawadogo<sup>2</sup>

<sup>1</sup>Department of Dentistry and Endodontics, Odonto-Stomatology Faculty, Felix Houphouët Boigny University, UFHB, Abidjan, Ivory Coast

<sup>2</sup>Faculty of Pharmaceutical Sciences of Felix Houphouët Boigny University UFHB Abidjan, Abidjan, Ivory Coast

**Objective:** The objective of this study was to assess the oral state of patients with homozygous sickle cell disease (SSD) to identify preventive and curative care needs.

**Methods:** Our study was carried out in the first Clinical Hematology service in West Africa in the Yopougon University Hospital (Abidjan- Côte d'Ivoire). A retrospective analysis of SS patients' records followed by interviews and dental screenings was carried out. The data collected was processed with the use of EPI Info software version 6.01.

**Results:** Sixty patients were examined. Detected pathologies were chronic pulpitis (33%), dentinal sensitivity (23%), chronic apical periodontitis (17%), and pulpal necrosis (10%). Dental care needs were surgical (14%), prosthetic (16%), conservative (24%) and prophylactic (46%).

**Conclusion:** The importance of this work is to propose a dental management of patients with homozygous sickle cell disease.

**Keywords:** Dental manifestations; dental treatment; homozygous sickle cell.

Sickle cell (SS) is a hemoglobinopathy resulting in an anemia due to the distortion of red blood cells (RBCs) into sickle-shaped. This condition is common with mass migration and intermingling of populations and is particularly widespread in black African and Indian populations. It is, for example, the most common genetic disease in France. The prevalence of hemoglobin S is 12% in Côte d'Ivoire.

Oral complications of the sickle cell disease (SSD) have been established since the end of the last century's first clinical cases. Oral signs of SSD include oro-facial pain, mesiofacial overgrowth, malocclusions, enamel hypominer-

alization and a caries-promoting factor.<sup>[1,2]</sup> Indeed, this is due to the sickle erythrocyte that upsets the blood flow in the pulp environment. In addition to orofacial syndromes, the malocclusions were aggravated by the association of the hypoxia to the jaw osteomyelitis and the bone marrow hyperplasia.<sup>[3,4]</sup> In the dental environment, the major form of the disease characterizes the described pathophysiology. It is the homozygous SSD, defined as a chronic hemolytic anemia interspersed with hemolytic seizures and vaso-occlusive attacks often complicated by severe bacterial infections.<sup>[5,6]</sup> A pulpal necrosis caused by the association of hypoxia with decreased blood supply to the teeth disorder

**Correspondence:** Dr. Yolande Koffi-Gnagne. Felix Houphouët Boigny University Ufhabidjan, 22 Bp 612 Abidjan 22. 612 Abidjan - Cote d'Ivoire.

Tel: +225 – 0110 8082 e-mail: gkyolande@gmail.com

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affected dental tissues.<sup>[2]</sup> Indeed, the erythrocytes sickle-shaped impedes the pulp microcirculation. Recent studies have also shown that the oral environment of children suffering from the pathology is an enabling environment of the spread of decaying disease pathogens.<sup>[7,8]</sup> Moreover, people's negligence regarding their oral health could lead to the handicap caused by hemoglobinopathies. All these factors combined to justify the poor oral state of patients suffering from this type of pathology. In Côte d'Ivoire, the prevalence of hemoglobin S is estimated at 12%, a similar prevalence is also found in other sub-Saharan countries.<sup>[9]</sup> Although Côte d'Ivoire shelters the first clinical hematology service in West Africa, no studies dealt with the oral health of SSD patients to ensure their full management. This study was therefore carried out to assess the oral state of patients suffering from homozygous SSD to identify preventive and curative care needs.

## Materials and methods

It was a two-month prospective study in the Clinical Hematology Department and at the Central Laboratory of the Reference Center. It was carried out in two phases involving a retrospective analysis of patients' records, followed by a prospective investigation combining interviews and oral examinations.

### Prospective survey

Medical records of hemoglobin phenotypic patients were consulted. Information on hematology analyses has been identified. The blood count included several parameters, some of which are the subject of our study. We specifically focused on (RBC) count. Values, which were admitted as a norm are typical [4-4.5] 106/MM3 in women and [4.5-6] 106/MM3 in men. Finally, data related to periodicity and type of attacks (hemolytic or vaso-occlusion) were recorded.

### Interviews and oral examinations

Selected patients were informed about the objectives of the study to obtain their informed consent to participate in the study. They were interviewed based on the WHO-1997 form of the annual frequency of SSD crisis and the association of dental pain with the onset of these attacks. Thereafter, an operator aided by an Assistant carried out clinical examinations. Plaque and dental tartar presence was used to determine every patient's Simplified Oral Hygiene Index (SOHI). Decayed teeth (D), missing due to extraction (M) and filled (F), were identified to calculate the average DMF index. Patients were then divided into two variables: (1) with cavities or (2) without cavities. Spe-

cial examinations of teeth were conducted on a cavity random basis, pulp vitality test (dental cold spray, Maccolux®, Luxembourg) and periodontal test (axial and transverse percussion). The standard practice of panoramic X-rays of the maxilla was done when needed with periapical X-rays (Kodak 6100®, Kodak, Paris, France).

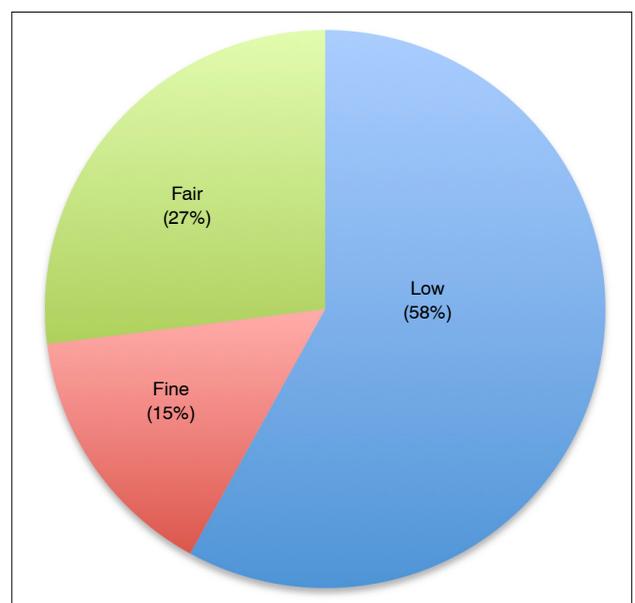
For each patient, the most severe dental pathology was selected as the only identifier assigned to the subject for statistical analysis.

### Statistical analysis

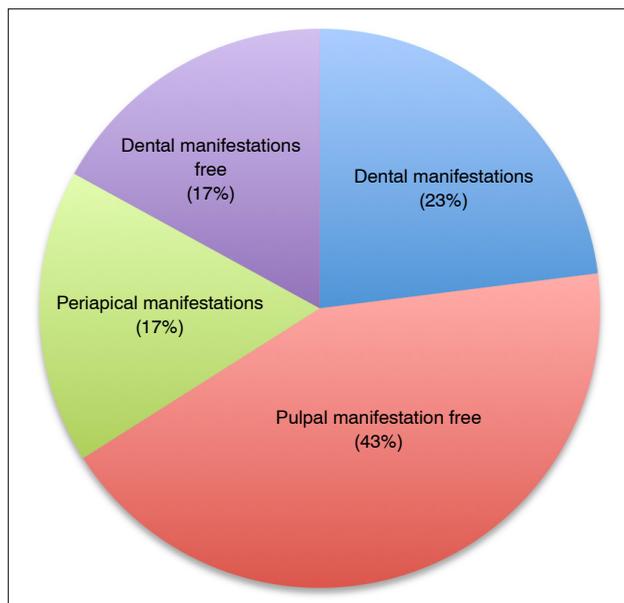
The data collected was processed with EPI Info software version 6.01. The statistical analyses were carried out by the KHI2 test and the Kruskal-Wallis non-parametric test was used to compare qualitative and quantitative variables averages. The significance threshold was set at 5% for observed differences.

## Results

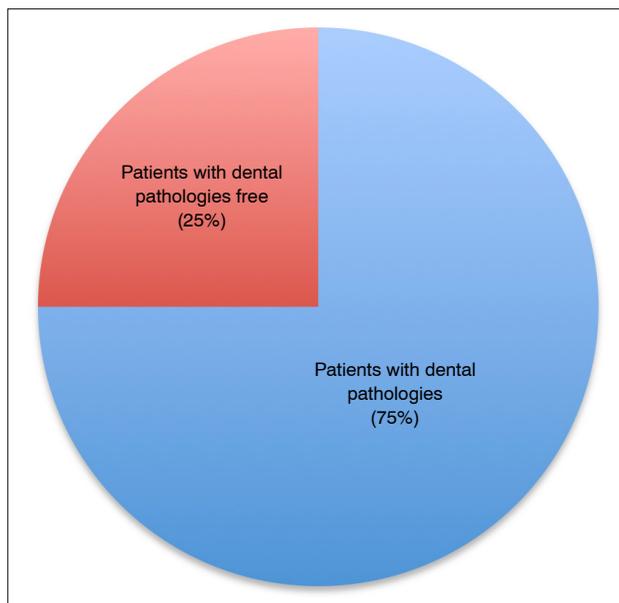
Sixty patients aged 12 to 40 years (average age = 20 years) made up the study sample. Gender-related distribution was 56.7% of women for 43.3%, giving a sex ratio of 0.7. More than half of examined patients (58%) had poor oral hygiene measured by the SOHIS (Fig. 1). The average DMF index was 3.25, reflecting the majority of carious lesions in 65% of tested patients. Patients had 83.3% of substance loss due to carious lesions or hypomineralization, divided into pulp attacks (43%), dentin attacks (23%) and periapical attacks (17%) (Fig. 2). Diagnoses of those attacks showed that chronic pulpitis was one-third of



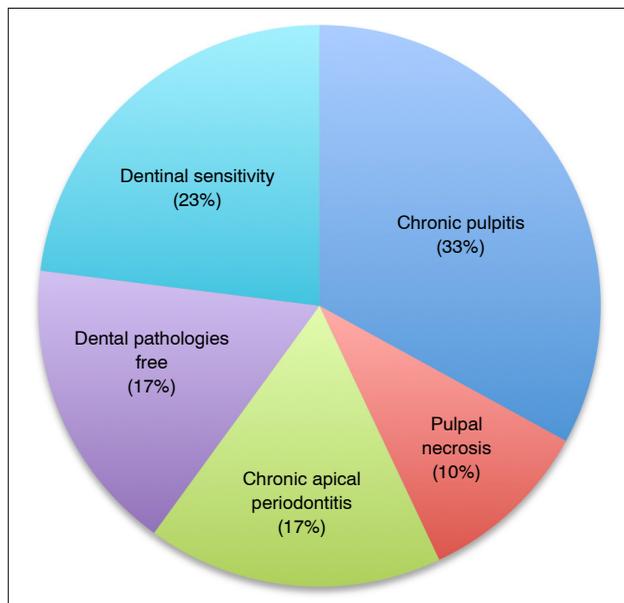
**Fig. 1.** Distribution of the SOHI in the study population. The majority of patients examined have unsatisfactory oral hygiene.



**Fig. 2.** Distribution of dental signs encountered in patients with sickle cell. Pulp inflammations with painful symptomatology are the most experienced manifestations of patients.

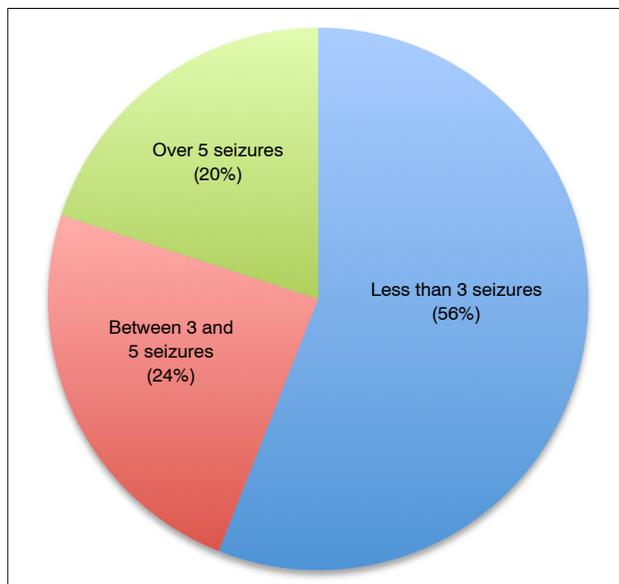


**Fig. 4.** Distribution of patients according to the occurrence of sickle cell attacks. 75% of patients with SSD seizures have cavities.



**Fig. 3.** Distribution of dental pathologies encountered in patients with sickle cell anemia. 1/3 of diagnosed teeth has chronic pulpitis.

identified pathologies (33%), followed by reversible pulpitis (23%), chronic apical periodontitis (17%), and pulpal necrosis (10%) (Fig. 3). 60% of teeth with healthy-looking and without loss of substance had chronic pulpitis. Some patients recall an episode of dental pain during their hemolytic or vaso-occlusive seizures. Said combination concerned 75% of patients with dental pathologies (Fig. 4) and the recurrence of these seizures was associated with a gradual disappearance of the pain (Fig. 5). Analysis of

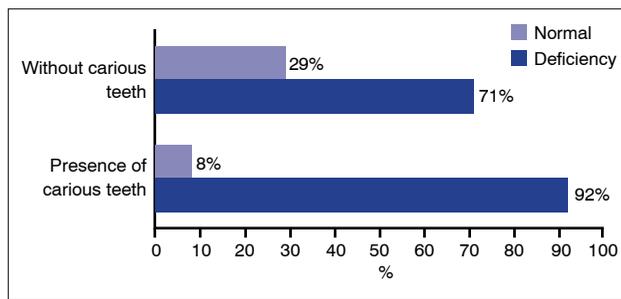


**Fig. 5.** Distribution of the occurrence of dental pain according to the annual frequency of SSD seizures. The more attacks are recurrent the less teeth are affected by the painful symptomatology ( $p=0.001523$ ).

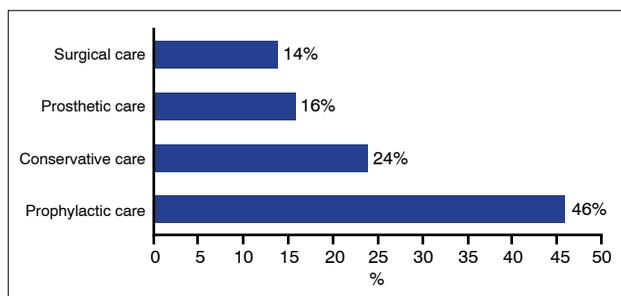
erythrocyte parameters revealed that 92% of patients with decayed teeth had RBC deficiency (Fig. 6). These data reflect, in ascending order, care needs: surgical (14%), prosthetics (16%), preservatives (24%), and caries disease prophylaxis (46%) (Fig. 7).

## Discussion

SS or SS anemia is the most common hemoglobinopathy, and predominant mainly at the geographical band between



**Fig. 6.** Distribution of patients according to red blood cell deficiency. The erythrocyte deficiency determines the sickle disease ( $p=0.0074$ ).



**Fig. 7.** Distribution of patients with sickle cell disease dental care needs. All patients examined need primary care.

the 15<sup>th</sup> parallel north and the 20<sup>th</sup> parallel south called “sickle belt” as defined by Lehmann.<sup>[5]</sup> This study shows that SSD has a wide variety of disorders at the dental organ by attacks on the enamel, the dentin, the pulp and the periodontium.

### Oral status of patients with sickle cell disease

As described by early works about SS dental symptoms, our study shows that the dental pulp is the most affected tissue by the disease.<sup>[11]</sup> The Vaso-occlusive crisis characterizes the critical phase of SSD and first clinical signs are pains in the child’s limbs and in the adolescent’s abdomen.<sup>[12,13]</sup> Tested patients from this study had dental pains during vaso-occlusive and hemolytic attacks. The pulp vaso-occlusive crisis causes an ischemia which describes the dental pain etiology.<sup>[5]</sup> First studies in Côte d’Ivoire had focused on the screening and medical management of the disease.<sup>[10]</sup> This study highlights the frequency of pulp and periapical chronic pathologies. Firstly, the high prevalence of chronic pulpitis is due to the sickle cell in the dental pulp during pulp and calcifications attacks.<sup>[11,14,15]</sup> Secondly, the evolution towards the necrosis represents a bacterial source capable of initiating apical periodontitis. By a considerable number, reversible pulpitis related to dentin lesions is the second detected dental pathology. It was proven that the SS patient oral environment’s alterations promote the initiation and hosting of amelo-dentin lesions. Indeed, children

with SS have a reduction of the salivary buffering capacity<sup>[7]</sup> and a greater virulence of pathogen bacteria associated with carious lesions,<sup>[8]</sup> due to an increase in production of acids which lower the pH of the buccal environment. In this context, the hypomineralization of hardened tissue in SS patients is an extra favorable setting for the loss of dental substance in the event of a bacterial attack.

This investigation also reveals that patients with carious lesions have a greater recurrence of SSD’s seizures. This recurrence of crises is associated in the long term with the loss of the tooth vitality. Hemolysis in the dental pulp also helps to install asymptomatic irreversible pulpitis without the loss of clinically perceptible hard tissue. Observation of erythrocyte parameters revealed that patients with low RBC count are more prone to caries. Indeed, because these cells are blood iron vectors, current studies show that iron deficiency anemia increases the susceptibility for developing a carious disease.<sup>[16,17]</sup>

### Dental treatment needs

Analysis of results showed that the high DMF index of the study population was due to the frequency of decayed teeth and missing teeth. A need for prophylactic care is, therefore, necessary for all seen patients. This concerns the detected enamel and dentin initial lesions. However, preventive care is necessary for all patients. The correlation between the recurrence of SSD attacks and the presence of necrotic teeth is proven.<sup>[18]</sup> Our results show that major curative treatments concern endodontic therapy. Identified chronic pathologies also showed the frequency of teeth in their root state, requiring avulsions. This could justify the need for occluso-functional rehabilitation in these patients.

From an epidemiological point of view, these dental care needs are not specific to SSD patients. But it is about a population in need of special medical care, not to exacerbate or deteriorate their general health status. The orofacial sphere is not the only one affected by manifestations of SS, but once the diagnosis of the disease has been established, oral rehabilitation and the application of preventive measures should be introduced. In this sense, the RBC can be a first indicator of the introduction of the dentistry therapeutic strategy and the treatment of oral complications must be adapted to each patient’s systemic state and special needs.

### Conclusion

This study complements existing data on SS anemia’s oral consequences. For obvious reasons, emphasis must be placed on a multidisciplinary approach in the monitoring of patients with SSD. The objective of this work is to sug-

gest a dental management of patients with homozygous SS. It goes through the integration of a systematic oral examination from the presumption of the disease to the introduction of preventive measures adapted to each patient, and the supervision for the treatment of oral complications due to the recurrence of SS attacks.

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