ORIGINAL ARTICLE

Dental and periodontal health status of subjects with sickle cell disease[†]

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Abstract   *Background/purpose:* Sickle cell disease (SCD) is a chronic, hereditary, autosomal recessive disorder. The pathophysiology of SCD is thought to result from polymerization of hemoglobin S in red blood cells under hypoxic conditions, which results in vaso-occlusion. The aim of this study was to determine the periodontal and dental health status of patients with SCD.

Materials and methods: Fifty-five SCD patients and 41 healthy individuals were evaluated. Detailed medical and dental histories were taken, and a record made of dental status (missing teeth, restorations, impacted teeth, root canal treatment), periodontal status [plaque index (PI), gingival index (GI), probing depth (PD), bleeding on probing (BOP)], alveolar bone level (ABL), mandibular cortex index, and bone quality index.

Results: Two hundred and six teeth were missing, and a total of 195 teeth had restorations. Between-group differences existed for the PI, GI, and BOP; these variables were higher in patients than in the healthy individuals ($P < 0.0001$). No between-group differences existed for PD. In patients, there was a positive correlation between PD and BOP ($P < 0.0001$; $r = 0.657$), PD and GI ($P = 0.02$; $r = 0.299$), PD and PI ($P = 0.01$; $r = 0.343$); BOP and GI ($P < 0.0001$; $r = 0.503$), BOP and PI ($P < 0.0001$; $r = 0.496$); and GI and PI ($P = 0.003$; $r = 0.388$). The ABL in patients was found to be similar to that of the general population with an unknown periodontal condition.

Conclusion: No clinical periodontal disease or attachment loss was detected in patients. However the PI, GI, and BOP were significantly higher in patients with SCD, which may reflect an as yet undefined variable response to microbes. There were no significant differences,

however, in pocket depth between the two groups. Therefore we are unable to confirm any significant relationship between SCD and periodontal diseases. Oral health is not a major concern for SCD patients. The reason for this finding may be the potentially severe complications of SCD, which mean that oral and dental problems are not major concerns for this particular group of patients.

Introduction

Sickle cell disease (SCD) is a hereditary, autosomal recessive disorder. This chronic disorder has been linked to hypoxia. The pathophysiology of SCD is thought to result from polymerization of hemoglobin S in red blood cells (RBCs) under hypoxic conditions, which results in the occlusion of blood vessels. The single amino-acid change in the beta subunit causes sickle hemoglobin to polymerize, especially under low oxygen tension. Polymerization causes the RBCs to deform into the characteristic sickle shape, thus plugging blood vessels.[1] A decrease in the tissue pH and oxygen tension leads to further sickling. When hemoglobin S accounts for <40% of the patient's hemoglobin, sickling occurs only under conditions of severe hypoxia.

The common clinical manifestations of SCD are vaso-occlusive crises, avascular necrosis (AVN), and osteomyelitis.[2] Hemoglobin S polymerization can lead to hemolysis, inflammation, cell adhesion, ischemia-reperfusion injury, infarction, endothelial dysfunction, increased superoxide production, and characteristic vaso-occlusive crises resulting from acute vascular obstruction and vascular dysfunction.[3]

A sickle cell crisis may be precipitated by dehydration, acidosis, trauma, overexertion, surgery (including tooth extraction), infections, general anesthesia, temperature extremes, vascular occlusion, and pulmonary disease. Patients may have fatigue and weakness; pain in the long bones, joints, and abdomen; cardiomegaly; systolic murmur; epistaxis; icteric sclera; bone deformities; and pallor of the oral mucosa.[4] Problems in the cardiopulmonary system, kidneys, bone, and central nervous system, and ophthalmic complications develop secondary to damage caused by vaso-occlusive episodes in the respective capillary networks.

Patients with SCD are more prone to infections because of hyposplenism, which occurs as a result of repeated infarctions leading to atrophy and an autosplenectomy. Impaired splenic functions lead to impaired removal of microorganisms from the blood. Changes in body temperature and tissue pH that occur as a result of infection may precipitate a vaso-occlusive crisis.

Although oral problems were described in the literature, these are less common than other medical problems. Oral problems such as unilateral infarct of the mandible,[5,6] pulpal necrosis,[5,7] osteonecrosis,[8] facial swelling,[1] diastema, hypodontia,[9] gingival enlargement,[1] palatal pallor,[4] increased risk for caries,[10] osteomyelitis of the mandible,[11] unilateral anesthesia,[6,8] midfacial overgrowth,[4] and orofacial pain[4,11,12] were reported.[13]

Findings on panoramic radiographs may include an enlarged medullary space, a stepladder-like effect on the bone, enamel hypomineralization, calcified pulp canals, an increased prevalence of mandibular osteomyelitis, and thinning of the inferior border of the mandible.[4,8,13,14] Digital analysis of the trabecular pattern in the jaws of patients with SCD revealed a less complex trabecular structure in both the maxilla and the mandible.[15,16]

The aim of this paper is to present dental and periodontal views of patients with SCD who had been sent to the periodontology department for a consultation. This paper also presents current knowledge and suggestions for SCD patients in dental practice.

Materials and methods

The study protocol was approved by the Ethics Committee of the Medical Faculty of Baskent University, in accordance with the Declaration of Helsinki (D-KA06/04). SCD patients are regularly evaluated at the hematology clinics of Baskent University. They are sent to the periodontology department for dental and periodontal examinations and consultations as part of a standard medical examination. Written, informed consent was obtained from all participants before their examination. Participants' genotypes were confirmed by electrophoresis. Complete medical and dental histories were noted, radiographic examinations were done, and data regarding smoking habits, consanguineous marriages, previous surgeries (especially a splenectomy), and a record made of the existence of osteoporosis, hepatitis, and physical disabilities.

The plaque index (PI; Silness and Löe) and gingival index (GI; Löe and Silness) were scored; probing depth (PD) was measured in millimeters with a Williams periodontal probe (Carl Martin, Solingen, Germany). Bleeding on probing (BOP) was recorded as absent or present from four aspects (mesial, buccal, distal, and palatinal/lingual) of all existing teeth, with the exception of third molars, in the mouths of all subjects.

Fillings, missing teeth, impacted teeth, tooth canal treatments, and periapical lesions were recorded. All panoramic radiographs were taken by the same experienced radiograph technician, and assessed by an experienced periodontist.

Mandibular cortex widths, the mandibular cortex index and bone quality index, and alveolar bone level were recorded from the panoramic radiographs.

Mandibular cortex widths were measured as the height of the inferior cortex on the left side of the mandible divided by the height of the lower border of the mandible to the edge of the mental foramen.[17–19] The mandibular cortex index describes the appearances of the lower

mandibular inferior cortical shape as smooth (C1), with semilunar erosions (C2), or as porous, severely eroded (C3).[18,19]

Bone quality was assessed by a bone quality index.[18] The bone quality index describes bone quality on the basis of the amount and proportion of cortical and trabecular bone; the four classes are: homogenous cortical bone; thick cortical bone with a marrow cavity; thin cortical bone with dense trabecular bone of good strength; and very thin cortical bone with low-density trabecular bone of poor strength.[18]

The alveolar bone level was measured with a Björn and Holmberg ruler,[20] which had previously been used by our group.[21,22] This transparent ruler has six divisions, and determines the height of the interdental alveolar bone as a proportion of the total length of the tooth. The ruler was applied over the radiographs, with the coronal baseline falling over the incisal edge or crown tip of the tooth to be measured. The inclination of the coronal baseline toward the image is essential. The mesial and distal aspects of each tooth, with the exception of the third molars, were individually measured and recorded. The optimal bone height was considered to be 65% of the total tooth length. This "optimal bone height" was divided into four equal portions, scored as 1, 2, 3, and 4, and a score of "0" indicates 65% $\pm$ 5% of the total root length. The scoring system is summarized as follows: $0 =$ no or initial marginal bone loss; $1 = <25\%$ of the total tooth length lost; $2 = <50\%$ and $>25\%$ of the total tooth length lost; $3 = <75\%$ and $>50\%$ of the total tooth length lost; and $4 = >75\%$ (severe) alveolar bone loss.

The radiographic data obtained from patients with SCD were compared to data obtained from patients with an unknown periodontal condition. Detailed information about this population was previously documented in detail.[21,23,24]

Results

Demographic evaluations

We clinically evaluated 55 SCD patients (30 women and 25 men; mean age 31.1 ± 9.7 years) with homozygous SCD in a steady-state condition and 41 healthy, age- and sex-matched controls (25 women and 16 men; mean age 27.8 ± 6.3 years) with no history of SCD; the panoramic radiographs of 54 SCD patients were evaluated. No significant between-group differences were recorded with regard to age, sex, or smoking habits (Table 1). Consanguineous marriages existed in 31.4% of the SCD patients.

Medical evaluation

Forty-nine percent of SCD patients had at least one another medical problem; these problems included complications of SCD such as cardiovascular diseases, ophthalmic disorders, and long-bone pain, and 40.4% of them had avascular necrosis in their hips and scapulas (Table 2).

No significant differences existed between the number of erythrocyte transfusions and the rate of having a crisis in the previous 3 years.

During our research, while neither periodontal infections nor abscesses contributed to vaso-occlusive crises, two patients were sent to the dental clinic for consultation soon after control of a vaso-occlusive crisis. Both patients had deep caries and periapical lesions with submandibular swelling.

Periodontal and dental evaluations

Significant between-group differences existed for PI scores ($P < 0.0001$), GI scores ($P < 0.0001$), and BOP positive sides ($P < 0.0001$). These variables were higher in the SCD group. No between-group differences existed for PD ($P > 0.05$) (Table 3).

In the SCD group, positive correlations were found for PD, PI, GI, and BOP; and between PD and BOP ($P < 0.0001$; $r = 0.657$), PD and GI ($P = 0.02$; $r = 0.299$), PD and PI ($P = 0.01$; $r = 0.343$), BOP and GI ($P < 0.0001$; $r = 0.503$), BOP and PI ($P < 0.0001$; $r = 0.496$), and GI and PI ($P = 0.003$; $r = 0.388$) (Table 4). In the controls, only GI was correlated with PD ($P = 0.01$; $r = 0.370$) and BOP ($P < 0.0001$; $r = 0.695$) (Table 5).

Two hundred and six teeth were missing, and 44.66% of these were first and second molars. There were 105 fillings (7.77% of the existing teeth), 90 crowns or bridges (6.47%), and 11 teeth which were beyond repair (0.79%) detected (Table 6).

Evaluation of panoramic radiographs

Fifty-four panoramic radiographs belonging to SCD patients were evaluated for dental aspects (periapical lesions, impacted and missing teeth, root canal treatment, and the mandibular cortex index). One panoramic radiograph could

Table 1 Epidemiologic data regarding to age, sex, and smoking habits.

	SCD	Control	P
Sex			
Female	30	25	NS
Male	25	16	
Age (years)	31.1 ± 9.7	27.8 ± 6.3	NS
Consanguinous marriages	31.4%	N/A	
Smoking	5.9%	2.4%	NS

SCD = sickle cell disease.

Table 2 Medical health condition of patients with sickle cell disease (SCD).

Avascular necrosis	40.4%
Systemic disease (at least one other than SCD)	49%
Hepatitis history	32%
Erythrocyte transfusion (in past 2 y)	44.4%
Having crisis (in past 2 y)	78.3%
Hospitalization (in past 2 y)	71.8%

Table 3 Periodontal parameters in patients with sickle cell disease (SCD) and controls.

Periodontal parameters	SCD	Controls	P
Plaque index (mean ± ss)	1.81 ± 0.78 (median = 1.96)	0.19 ± 0.17 (median = 0.14)	<0.0001
Gingival index (mean ± ss)	0.46 ± 0.39 (median = 0.34)	0.16 ± 0.12 (median = 0.14)	<0.0001
Probing depth (mean ± ss)	1.94 ± 0.5 (median = 1.8)	1.94 ± 0.2 (median = 1.97)	NS
Bleeding on probing (% ± ss)	26.6 ± 19.1 (median = 23.0)	6.4 ± 4.8 (median = 5.0)	<0.0001

not be evaluated because it was kept by the patient at his request.

Thirty-nine root canal treatments were detected in 54 patients. Of these treated teeth, 90% had radiolucent areas or a widening of the periodontal ligament in the periapical region without clinical symptoms.

Forty periapical lesions (2.88%), 39 root canal treatments (2.81%), 11 teeth beyond repair, and 24 impacted teeth were seen (Table 6).

A stepladder-like effect was seen at interradicular sites in 23.7% of patients. Osteoporotic changes were detected in the body of the mandible in 37.1% of SCD patients. The mean mandibular cortex width was calculated to be 4.32 mm. The distribution of scores was found to be 26.4%, 45.3%, and 28.3% for the mandibular cortex index, and 5.7%, 7.5%, 35.8%, and 50.9% for the bone quality index in patients with SCD.

Alveolar bone levels at mesial and distal sites are shown in Table 7. A score of 1 was the most frequent score at both mesial and distal sites in all cases.

Discussion

SCD was first described in 1910, in a black West Indian dental student who presented with pulmonary symptoms, and the physical and chemical properties of hemoglobin of individuals with SCD were identified by Pauling et al.[25]

SCD was first recognized in people of West African ancestry. Hemoglobin S occurs with the greatest prevalence in tropical Africa; the heterozygote frequency is usually about 20%, but in some areas reaches 40%. The sickle cell trait has a frequency of about 8% in African–American populations. The hemoglobin S gene is found to a lesser extent in the Middle East, northern Greece, countries bordering the Mediterranean (Sicily and other parts of southern Italy, Turkey along the southeastern coast, and Saudi Arabia, especially the eastern province), the African coast, and in aboriginal tribes in India.[3,26] On occasion, SCD is found in people of European extraction, especially when racial admixture has occurred over the centuries.[3]

SCD may be diagnosed in the 16th week of gestation, but manifestations do not appear until the 6th month after birth.[27] The National Cooperative Study of Sickle Cell Disease estimated that the median survival of individuals with SCD is 42 years for men and 48 years for women.[28] The mean age of examined patients with SCD in the present study was 31 years. Platt et al[29] reported the death of patients with SCD over the age of 20 years. During a 2-year period, two of 55 patients who were examined in our hematology clinic died; they were males, aged 20 and 24 years respectively. The reason for the low number of deaths may be related to the peak incidence of death that occurs between 1 and 3 years of age (deaths among patients younger than 20 years are predominantly caused by pneumococcal sepsis) or to our study cohort being well maintained patients. Although men are at greater risk of early death than women, there was no evidence that the risk factors for early death differed according to sex in SCD patients.[28]

Pulmonary hypertension, acute chest syndrome, and sepsis have been reported as reasons for sudden death in adult patients.[28,30] Another sudden death risk arises from excessive exercise; reduction in pH and temperature at tissue level facilitates oxygen delivery during exercise, and these changes lead to higher concentrations of deoxygenated hemoglobin.[31] Infections and sepsis remain the most common causes of death and vaso-occlusive crises for patients with SCD, and dental or periodontal infections[4] may play a part in precipitating a crisis.

There is no cure for SCD, and management of this disease is usually unsatisfactory. The prognosis of SCD patients is not known; however, individuals with SCD have a severe clinical course with acute pain and often require hospitalization for vaso-occlusive crises and other complications of the disease.[29,30] Although all of the patients evaluated in our clinic were well maintained by our hematology clinic, 72% of them had been hospitalized during the past 2 years, and the mean number of days spent in hospital per year was 28 days. Seventy-eight percent of these patients had a mean of seven crises per person during the past 2 years. Sickling is undoubtedly an important

Table 4 Correlations between periodontal parameters in patients with sickle cell disease (SCD).

SCD group	PI	GI	PD	BOP
PI				
GI	P = 0.003; r = 0.388			
PD	P = 0.01, r = 0.343	P = 0.02, r = 0.299		
BOP	P < 0.0001, r = 0.496	P < 0.0001, r = 0.503	P < 0.0001; r = 0.657	

BOP = bleeding on probing; GI = gingival index; PD = probing depth; PI = plaque index.

Table 5 Correlations between periodontal parameters in control group.

Control group	PI	GI	PD	BOP
PI		NS	NS	NS
GI			P = 0.01; r = 0.370	P < 0.0001; r = 0.695
PD				NS
BOP				

BOP = bleeding on probing; GI = gingival index; PD = probing depth; PI = plaque index.

Table 7 Alveolar bone level (ABL) scores according to mesial and distal sites for all patients.

ABL Scores	ABL − mesial	ABL − distal
0	33.3	21.3
1	52.7	54.1
2	11.2	20.6
3	1.2	2.4
4	0.3	0.5
Undecipherable	1.3	1.2

accelerator of organ destruction and organ failure; this could explain why 49% of our patients had another systemic disease other than SCD.

In the present study, the plaque and gingival indices were significantly higher in SCD patients than in healthy individuals. Usually, oral health is not a primary concern of SCD patients. In the current evaluation, no significant differences existed regarding the PD between patients with SCD and healthy individuals. These results agree with the findings of Crawford[32] and Arowojolu and Savage.[33] Crawford[32] suggested that SCD is not associated with increased levels of gingivitis and periodontitis in patients with SCD. In agreement with the results of a study by Crawford,[32] Arowojolu and Savage[33] found no significant difference in alveolar bone loss patterns between patients and controls. In the same study group, Arowojolu[34] found no clinical periodontal disease or attachment loss in patients with SCD. However, in the present study, the number of BOP-positive sites was greater in SCD patients than in controls, but there was no clinical attachment loss. Clinically, gingival tissues in SCD patients in our group appeared faded and yellowish, which is compatible with previous reports.

SCD has long been recognized as an inflammatory condition. This ongoing inflammatory state seems to contribute to sickle cell complications. In the present study, patients were examined when they were in a steady-state condition. Steady-state disease does not necessarily mean an absence of ongoing pathophysiologic activity, as shown by the elevation and fluctuation of acute-phase reactants and cytokines in patients with steady-state SCD.[35−37] High levels of endothelial progenitor cells in the circulation can indicate endothelial activation and

inflammation.[38] In another study, human subjects with and without periodontal disease were cross-sectionally examined.[39,40] Gingival inflammation directly correlated with the level of periodontal disease, the level of putative pathogens, and the level of short-chain carboxylic acids (SCCAs).[40] SCCAs are metabolic intermediates that occur in and are released by bacteria and animal cells. On a site basis, however, when probing depth, tooth location, and bacteria were controlled for, gingival inflammation was significantly correlated only with the concentrations of propionic and butyric acids, not with the level of any single putative pathogen.[39] These experimental findings support the hypotheses that bacterially derived SCCAs are associated with human gingival inflammation, are not associated with gingival health, decrease following therapy, and can elicit a clinically defined gingival inflammatory response.[41] SCCAs may stimulate an inflammatory response by increasing leukocyte recruitment, viability, and cytokine gene expression of neutrophils.[41] It is remarkable that SCCAs stimulate an inflammatory response when placed on healthy human gingiva. In addition to stimulating an inflammatory response, potential therapeutic uses for SCCAs would have been identified for SCD patients. Butyric acid is normally used as a food additive, and it may increase the amount of fetal hemoglobin in the blood. An intravenous infusion of butyric acid into patients with SCD drives the anemia into remission by downregulating sickle globin gene expression and upregulating fetal globin gene expression. It may be possible to begin to test and identify pharmaceutical agents which diminish the harmful aspects, while enhancing the beneficial components of the inflammatory response (for reviews, see Perrine et al,[42] Schlake et al,[43] Orkin et al,[44] and Niederman et al[41]).

Table 6 Evaluation of panoramic radiographs.

Teeth	Fillings	Prosthetic restorations	Root canal treatment	Periapical lesion	Teeth beyond repair	Impacted teeth	Missing teeth
Centrals	11	11	2	9		2	5
Laterals	6	11	1	4			5
Canines	2	17	8	4		1	
1st premolars	10	6	6	9	1		19
2nd premolars	10	18	6	13	3		29
1st molars	38	8	9	21	7		71
2nd molars	28	19	7	20			21
3rd molars						21	56
Total	105	90	39	40	11	24	206

Neutrophils and eosinophils are a potential source of mediation, and leukocyte-derived mediators contribute to the inflammatory response and in turn affect vascular tone and permeability. Mononuclear cells from SCD patients release more superoxide than do healthy controls when stimulated with 12-O-tetradecanoylphorbol-13-acetate (TPA), also commonly known as tetradecanoylphorbol acetate, tetradecanoyl phorbol acetate, and phorbol 12-myristate 13-acetate (PMA) and zymogen.[45] Nitric oxide is an important vasodilator. Superoxide inactivates nitric oxide and produces oxidant damage.[46] Dias-Da-Motta et al[45] suggested that this phenomenon could represent an additional risk factor for obstructing the microcirculation and causing tissue damage in SCD patients. Once oxidative damage occurs in RBCs, hemoglobin denatures, and Fe^{3+} interacts with the RBC membrane causing lipid peroxidation, membrane damage, inactivation of membrane enzymes, and DNA damage.[47] Ozdogu et al[48] demonstrated that apoptosis of blood leukocytes is increased in patients with SCD compared to controls. Conversely, few apoptotic cells are apparent in the inflammatory infiltrates of periodontitis tissue. Lard et al[49] showed that neutrophils are activated in SCD patients, suggesting an important role in the pathophysiology of SCD. The observed neutrophil activation in sickle cell patients may lead to increased adherence to the endothelium in the microcirculation. For instance, neutrophils in localized aggressive periodontitis are already primed (activated), and superoxide levels are increased. The result in patients with localized aggressive periodontitis is severe periodontal destruction that cannot be explained by bacterial invasion. Similarly, neutrophils in SCD patients are activated, and a high amount of plaque was observed. However, the clinical appearance was not compatible with these findings. We suggest that one of the reasons for this incompatibility (characteristic of patients with SCD) would be the vascular pathobiology.

The management of chronic pain in SCD is usually achieved by nonsteroidal anti-inflammatory agents (NSAIDs). Adjunctive therapeutic strategies that modulate inflammatory mediators can play a significant part in periodontal therapy. It was shown that the use of NSAIDs as an adjunct to nonsurgical management of chronic periodontitis caused a significant decrease in the probing depth.[50] The results of host modulation studies showed that this approach may be a potential target to reduce tissue destruction. NSAIDs block proinflammatory cytokines; a subdose of doxycycline blocks metalloproteinases, and bisphosphonates block osteoclast activity.[51,52] Adjunctive anti-inflammatory therapy may lead to a faster and greater reduction in probing pocket depth than conventional periodontal therapy.[53] So, the regular use of NSAIDs by SCD patients may result in prevention of periodontal destruction in those patients.

Patients with SCD are susceptible to overwhelming infection. Patients with SCD and unexplained fever should be thoroughly examined.[54] If the clinical condition suggests septicemia, broad-spectrum antibiotics should be prescribed. Continued use of antibiotics by SCD patients can control periodontal destruction.

Sometimes, the severity of anemia in patients with SCD gradually increases as they age. The reason for this marrow "burn-out" phenomenon is unknown. The clinical situation is complicated by the fact that many patients have organ damage, such as dilated cardiomyopathy, that may limit their ability to tolerate such severe anemia.[54] Also, the cumulative effects of periodontal diseases are apparent in older persons.[55] In the present study, the mean age of the SCD patients was 31 years; hence it was not surprising to detect less periodontal destruction in spite of the presence of higher plaque and gingival indices.

SCD complications place patients at risk for poor psychosocial adaptation, including depression and anxiety symptoms. Hijman et al[56] showed that children with SCD are at increased risk of developing severe internalizing problems as a result of their disease. Subgroups of children with SCD also appear to be at increased risk of developing severe externalizing problems, which may be related to either sociodemographic factors or disease factors, such as neurocognitive deficits associated with cerebral infarction. Children with SCD are also perceived to have more difficulties than healthy siblings in school functioning, demonstrate less-competent social behavior, and tend to have more attention deficits. Carpenter et al[57] reported that adolescents with SCD who classified themselves as having high behavioral inhibition displayed higher levels of anxiety and depression than adolescents with SCD who classified themselves as having low behavioral inhibition; adolescents with SCD in the middle behavioral inhibition category generally scored in between the other two categories.

Oral health is an integral and critical part of general health. Our observation that "oral health is not a priority for patients with sickle cell disease" is supported by the increased amount of plaque together with the higher number of missing teeth when considering the patients' ages in our study group. This may also explain why the mean PD was 1.94 mm despite higher plaque scores. Although all patients were offered free dental treatment and scaling and they were aware of the inadequacy of their oral health, only a few patients applied for treatment of their acute problems. The reason for this may have been severe complications of SCD. Another possible reason for less periodontal destruction would be patients' preference for tooth extraction in lieu of treatment. Treatments were provided to patients by experienced dentists and periodontists. Although oral hygiene did not reach satisfactory levels, no further dental or periodontal problems were detected. Hence, oral problems are not a major concern for this group of patients, as we have demonstrated. Patients' perceptions of the social and behavioral consequences of their oral conditions and their treatment likely have an important influence on their behavior toward oral health and their patterns of dental care utilization. Assessing patients' motivations and their desire to retain the natural dentition is an important component of a treatment plan. Most patients have complex medical conditions. In addition, it was reported that most patients experience depression, anxiety disorders, or somatoform disorders of which dentists need to be aware.

Acute bone problems in SCD were described by Almeida and Roberts[2] as vaso-occlusive crises, osteomyelitis, stress fractures, orbital compression, dental complications, vertebral collapse, and bone marrow necrosis; chronic bone problems were described as osteonecrosis, chronic arthritis, osteoporosis, and impaired growth. Most SCD

patients have joint and bone disease, especially pain caused by hip and scapular joint calcification, and 40.4% of the patients in our group had AVN in their hips and scapulas. Avascular necrosis or osteonecrosis occurs when vaso-occlusion results in an infarction of the articular surfaces and heads of the long bones.[2] Maxillofacial involvement, especially of both the maxillary and mandibular jaw bones, is estimated to be between 79% and 100% in SCD.[5,16]

In studies performed by Rohlin et al,[58,59] panoramic radiographs were found to be equivalent to periapical radiographs for the upper-jaw, but inferior to periapical radiographs for the lower jaw. As long as an ideal image quality can be achieved, panoramic radiographs can be used for dental and periodontal evaluations.

Previously, panoramic radiographs were used to identify postmenopausal women at risk of osteoporosis.[17,19,60,61] In panoramic radiographs, osteoporotic patients were shown to have a lower jaw bone mineral density than controls.[18] Brinker et al[62] showed an overall reduction in bone mineral density in children with SCD compared to normal subjects. Almost half of our SCD group had semilunar erosions in their mandibular cortex, and 87% of them had low-quality bone compatible with their medical history obtained from medical records. It is clear that panoramic radiographs are still useful diagnostic tools.

Routine treatment can be carried out during noncrisis periods; therapy during crises should be limited to pallia-tion. Ideally, a dental appointment should be scheduled in the morning with minimum treatment and a short visit. Antibiotic prophylaxis is recommended before all dental treatments. Local anesthetics with a vasoconstrictor can safely be used in these patients, because it is known that local circulation is adequate. Acetyl salicylic acid is not recommended because of the risk of inducing acidosis; acetaminophen and codeine combinations provide the best oral analgesia.[13,63] Dentists should be aware of dental pain of unknown cause in SCD patients. Dental pain in the absence of caries may be caused by an intrapulpal infarct, asymptomatic pulpal necrosis, or blockage of tiny pulpal vessels by sickle cell emboli.[7,13] If root canal treatment is not satisfactorily performed, an improperly filled tooth root may be a reason for a vaso-occlusive crisis because of inflammation.[8]

Conclusions

Appropriate dental and periodontal care improves a patient's quality of life by preventing eating difficulties, oral diseases, and esthetic concerns, and facilitates the management of the disease by the hematologist. Preventive dental care is essential for SCD patients.

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