Afr J Med Med Sci (2017) 46, 365-369

Occlusal presentations of individuals with sickle cell disease

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Abstract

Objective: To determine the occlusal presentations of individuals with sickle cell disease.

Materials and methods: One hundred and thirty five subjects with sickle cell anaemia and who had not previously received any form of orthodontic treatment were recruited from the sickle cell clinic/ Haematological out patient's clinics in four tertiary health care institutions in the south western part of Nigeria. Ethical approval was sought and gotten from each tertiary institution Ethical Committees involve in the study. All eligible subjects were seated on a comfortable chair had their oral examination done by a single author under natural lighting illumination for both dental intra – arch and occlusal parameters and all the data gotten were entered into a spread sheet and analyzed with SPSS version19 computer software. The level of confidence was set at $p < 0.05$.

Results: The age range was 10 - 49 years with majority of the sample falling into the age range 10 – 29 years. The sample comprised of 61 males and 74 females. Eighty nine percent of the samples were phenotypically positive for HbSS and also commoner among the female samples (57.0%). Occlusal anomalies such as increase overjet, anterior maxillary arch spacing and maxillary prognathism were observed in 40.7%, 58.5% and 45.2% respectively.

Conclusion: There is a high prevalence rate of malocclusion among HbSS individuals and they present with a variety of occlusal anomalies which will require the attention of the orthodontist in order to improve their aesthetics, function and psychological wellbeing.

Keywords: *Sickle cell anaemia, occlusion, orthodontics.*

Résumé

Objectif: Pour déterminer les présentations occlusales de personnes atteintes de drépanocytose.

Matériaux et méthodes: Cent trente-cinq sujets atteints d'anémie falciforme et qui n'avaient auparavant reçu aucune forme de traitement orthodontique ont été recrutés dans la clinique de drépanocytes/Cliniques hématologiques de patients non-hospitalisés dans quatre établissements de soins de santé tertiaires dans la région sud-ouest du Nigeria. L'approbation éthique a été cherchée et obtenue de chaquecomité d'éthique des institutions tertiairesparticipant à l'étude. Tous les sujets admissibles ont été assis sur une chaise confortable et ont eu leur examen oral par un seul auteur sous éclairage naturel pour les deux paramètres intracellulaires dentaires et occlusaux et toutes les données obtenues ont été entrées et analysées avec le logiciel SPSS version19. Le niveau de confidence a été fixé à $p < 0,05$.

Résultats: La tranche d'âge était de 10 à 49 ans avec la majorité de l'échantillon tombant dans la tranche d'âge de 10 à 29 ans. L'échantillon compris 61 hommes et 74 femmes. Quatre-vingt-neuf pourcent des échantillons étaient phénotypiquement positifs pour HbSS et aussi fréquent chez les femelles (57,0%). Des anomalies occlusales telles que l'augmentation du risque de surjet, l'espacement antérieur de l'arc maxillaire et le prognathisme maxillaire ont été observées respectivement dans 40,7% ; 58,5% et 45,2%.*Conclusion:* Il y a un taux de prévalence élevé de malocclusion chez les individus HbSS et ils présentent avec une variété d'anomalies occlusales qui nécessiteront l'attention de l'orthodontiste afin d'améliorer leur esthétique, fonction et bien-être psychologique.

Mots-clés: *Anémie falciforme, occlusion, orthodontique.*

Introduction

Sickle cell anaemia is a genetic disorder which results from point mutation of the beta chain of the haemoglobin gene resulting in abnormal haemoglobin variant referred to as haemoglobin S (HbS) [1]. This mutation causes the erythrocytes to change into sickle shape, which are destroyed more rapidly than normal red blood cells. Sickle cell anaemia results from the inheritance of the haemoglobin S gene from both parents, resulting in the homozygous state (HbSS) [2]. It is a hereditary and familial haemolytic disease, the most common genetic disorder amongst black people and one of the major chronic non-communicable diseases affecting man [3].

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Furthermore, the sickle-shaped haemoglobin loses their ability to undergo deformation which is required by them for navigating through the microcirculation. Hence, they cause obstruction in the capillaries bed and restrict blood flow to organs thereby resulting in ischemia, pain and often tissue damage.

It is the most common worldwide symptomatic haemoglobinopathy [4] and has an incidence of about 2% in Nigeria [5]. Although, the disorder has been documented in literature to be widely spread in Nigeria with a higher prevalence in the North (11.87%) [6]. Other variant of this disorder that also exists is the HbSC which occurs at a prevalence of 19% in the south and between 0.22 – 0.5 in the northern part of the country [6]

The prevalence of sickle cell anaemia in Nigeria is said to be on the increase, especially among the urban educated elite and in other communities with access to effective basic health care [7]. To compensate for the short life of the red blood cells, compensatory mechanisms associated with hyperplasia and expansion of bone marrow take place and these result in changes in bony structures. These changes can be observed on radiographs.

Studies have shown that individuals living with sickle cell anaemia may present with malocclusion in the form of maxillary gnathopathy [8], delayed tooth eruption and periodontitis [9], increased overjet and overbite [10]. Gnathopathy and excessive growth of the anterior part of the maxilla in the secondary dentition leads to prognathism, spacing and proclination of the incisor teeth [10]. These malocclusions results in psychosocial problems because of poor dental aesthetics, functional problems and susceptibility of teeth to trauma. It is believed that as the Nigerian population steadily grows, the likelihood of an exponential growth in the population of individuals with this disorder will be high hence many of them may likely seek orthodontic intervention to improve their smile, function and quality of life and self-esteem as a result of increasing awareness of the possibility of managing their occlusal discrepancies through the use of various orthodontic appliances.

This study therefore is aimed at determining the occlusal presentation of the sickle cell individuals of a Nigerian population with a view to ascertaining the level of their functional and psychosocial burden.

Materials and methods

The study was descriptive cross-sectional and carried out over a period of eight (8) months among consecutive sickle cell individuals aged 10 - 49 years attending sickle cell out clinics in four tertiary health care institutions in the south western part of Nigeria. The south western region of Nigeria is cosmopolitan and with various occupational groups hence, home to diverse ethnic groups and economic seat of Nigeria.

A total sample of 145 respondents was recruited out of which only 135 of them were analyzed due to incomplete data. The demographic record of each patient was retrieved from the case notes. Information not in the case note was either gotten from the patient, parents and or guardian. Ethical approval was sought and gotten from each tertiary institution Ethical Committees involved in the study (ADM/DCST/221/VOL.10, CHS/MMP/ERC/63, OOUTH/DA.326/T/7, 00005422). Also, verbal and written consent was sought and obtained from each subject involved in the study.

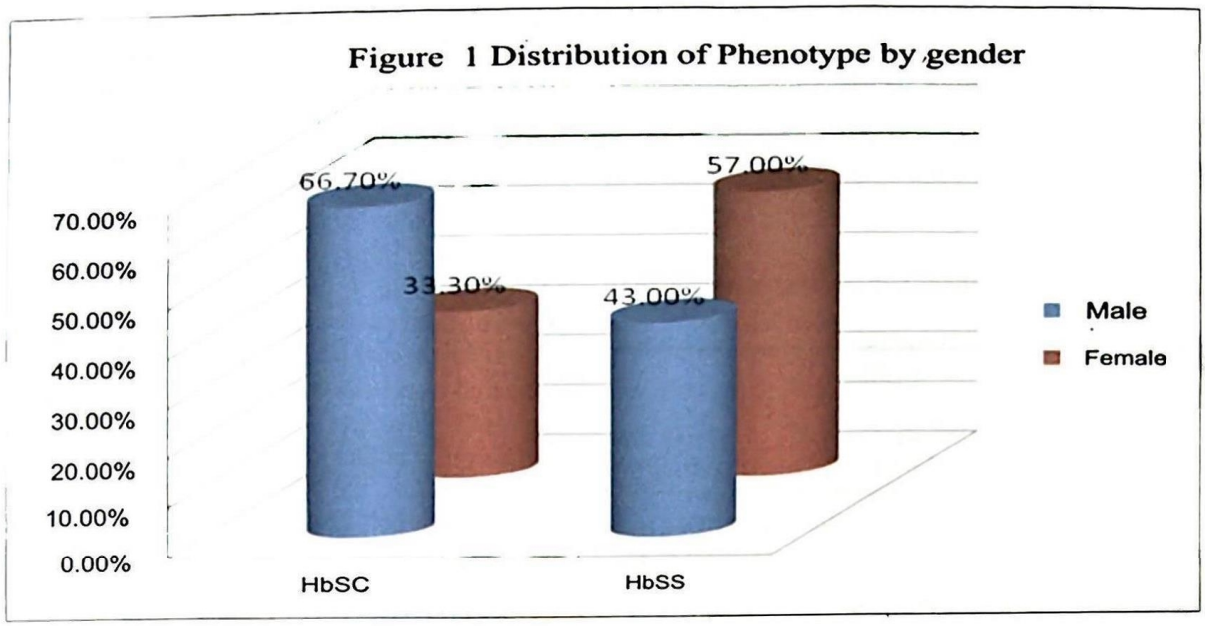
Inclusion criteria for subjects included; Subjects in steady health, no prior history of orthodontic treatment, full complement of teeth to avoid any confounding arch discrepancies as a result of tooth/teeth loss and an electrophoresis diagnosis of sickle cell anaemia. All eligible subjects had an oral examination done with gloved hands, facemask and a disposable dental explorer, dental mirror and a meter rule.

The information gotten was entered into a spreadsheet which consisted of two (2) sections. Section (A) recorded the demographic findings and the section (B) recorded the orthodontic oral findings.

Analysis was carried out using SPSS statistical software version 19. Data presentations were in the form of tables and charts (chi – square test and T - test was used to compare variables). An observation was considered to be statistically significant when the p value was less than 0.05.

Table 1: Demographic distribution of respondents

Age Range (years)	Gender		
	Female (count/%)	Male (count/%)	Total (count/%)
10 - 19	32	26	58
20 - 29	33	27	60
30 - 39	5	7	12
40 - 49	4	1	5
Total	74	61	135



Results

There were 135 samples examined and their ages ranged from 10 – 49 years. There were 61 males and 74 females with majority of the sample skewed around the age 10 – 29 years (Table 1). Only two main variants of the disorder were picked up during data collection and these were the HbSS (most common) and the HbSC variants. This study revealed that 89.0% of the samples were phenotypically positive for HbSS of which 57.0% of them were females while 11.0% were phenotypically positive for HbSC of which 66.7% of them were males. (Fig. 1)

Discussion

Nigeria is said to have the largest concentration of sickle cell anaemia individual worldwide [11, 12]. Numerous Nigerian studies have reported different prevalent rates for this genetic disorder which is put at between 2 - 2.9% [13, 14] in the southern part of the country to about 11.8% [6] in the north. Many of these studies are hospital based hence; the actual prevalence could be greater.

In this study, the most and least prevalent molar relations were Angle's Class I and Class III molar relationship which accounted for 61.5% and 11.1% respectively (Table 2). This is similar to a previous study in Lagos, Nigeria by daCosta *et al* [15] in 2005 among sickle cell individuals. Though, in their study they did not record any Angle's Class III molar relationship. Among the Angle's Class II molar relationship recorded in this study, it is very important to note that 97.3% of them presented

Table 2: Distribution of occlusal presentation in respondents

Occlusal presentation	Total count (%)
Anterior posterior relationship	
Angle's Class I	61.5
Angle's Class II	27.4
Angle's Class III	11.1
Asymmetrical molar relationships	0.04
Overjet	
Reduced/Reversed <2mm	11.9
Normal 2 - 4 mm	47.4
Increased >4mm	40.7
Mean overjet 3.96±2.15mm	
Overbite	
Anterior open bite (AOB)	8.1
Edge-Edge bite	8.9
Normal	73.3
Increased	9.6
Tooth:Arch relationshipUpper arch	
Anterior segmentNormal	17.8
Crowding	23.7
Spacing	58.5
Posterior segment	
Normal	80.7
Crowding	10.4
Spacing	8.9
Lower arch	
Anterior segment	
Normal	22.2
Crowding	34.8
Spacing	43.0
Posterior segment	
Normal	77.8
Crowding	16.3
spacing	5.9
Maxillary Prognathism	
Present	45.2
Absent	54.8

with Angle's Class II division 1 molar relationship. This might not be too surprising due to the presence of maxillary hyperplasia and expansion of bone marrow compensating for the short life span of erythrocytes [8, 10, 16] that is quite common in these individuals thereby often causing incompetence of the lips [8] and a proclination of the upper anterior teeth due to the lack of a lip seal [8].

Table 3: The incidence of malocclusion among respondents

Occlusion	Gender		
	Female count/%	Male (count/%)	Total (count/%)
Malocclusion	48(51.6)	45(48.4)	93(68.9)
Normal occlusion	26(61.9)	16(38.1)	42(31.1)
Total			135(100)

management for their malocclusion. This assertion had been previously suggested by Okafor et al.(17) who recommended in their study that sickle cell anaemia individuals should have access to orthodontic management associated with phonoaudiologic support in order to alleviate their conditions.

The tooth size arch length ratio discrepancies (arch spacing) recorded in this study were quite high and comparable to other previous studies in this group of individuals [15]. The arch spacing was more common and pronounced in the upper arch as compared to the lower arch. Maxillary hyperplasia as a result of haemopoeitic need might be responsible for this observation.

Therefore, the overall occlusal characteristics of the sickle cell individuals placed them with a malocclusion prevalence of 68.9% with no gender predilection (Table 3).

Table 4: Comparative analysis of occlusal characteristic with other studies

Occlusal presentations	*Present study%	*daCosta *et al* 2005 %	Onyeaso 2002 %	Sanu 2000 %	Isiekwe 1989 %
Angle's Class I	61.5	88.5	76.5	90.3	76.5
Angle's Class II	27.4	11.5	15.5	4.8	14.7
Angle's Class III	11.1	0	8.0	2.1	8.4
OverjetReduced	11.9	13.4	0.7	40.0	19.1
Increased	40.4	48.2	16.2	11.1	24.1
OverbiteReduced	17.0	7.6	1.4	7.3	5.2
Increased	9.6	21.2	3.8	15.6	13.5
Tooth:Arch Crowding (Upper)	23.5	10.6	-	14.6	25.8
Crowding (Lower)	34.6	-	-	25.3	41.9S
pacing (Upper)	58.	49.0	-	36.1	-
Spacing (Lower)	42.6	30.8	-	24.7	-

*Sickle cell population study. Others were on general population

Other occlusal traits observed in these individuals from this study included the highly increased overjet of 40.4% (Table 2). This is also similar to the observations by Taylor et al [10] and daCosta *et al* [15] in their study. The prevalence of increased overjet is quite alarming when compared with previous studies carried out among the general population though with an HbSS prevalence rate of about 2.9%. The implication of this is that the anterior teeth of these individuals are prone to a fracture and its complications usually following trauma due to their conspicuousness. In order to avoid these and the burden that might be associated with it which could affect the quality of life in this group of individuals, many might require orthodontic

Conclusion

Sickle cell individuals presents with a variety and high prevalence of occlusal anomalies which affect aesthetics, function, psychological and self-esteem. The malocclusion characteristics they present with further add to the burden they undergo as individuals due to their condition by reducing their oral health quality of life hence, it will be imperative that in order to reduce these burden physicians should refer them to dentist for dental education talk and a correction of their malocclusion.

Acknowledgment

The authors would like to thank the management of the Lagos University Teaching Hospital, Idi Araba,

Olabisi Onabanjo University Teaching Hospital, Sagamu, Ladoke Akintola University Teaching Hospital, Osogbo and the Obafemi Awolowo University Teaching Hospital Complex, Ile Ife for their support.

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