

WEST AFRICAN JOURNAL OF ORTHODONTICS

VOLUME 8, NUMBER 2

Print ISSN: 2315-9634
E-ISSN: 3141-5822

DECEMBER 2019

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Cell Anaemia**



**Tooth Movement in the resolution
of Crowding**



**Oral Ornaments and Orthodontic
Treatment**



Management of Midline Diastema



**Orthodontic Management of
Anterior Crowding**

Dental Arch Dimensions in Ghanaian patients with Sick Cell Anaemia

Siale EE^a, Newman-Nartey M^b, Amoah G^b

Abstract

Background: Sick Cell Anaemia (SCA) presents with multisystemic complications which may include dentofacial effects. Dental arch dimension variations have been demonstrated to occur in some chronic haematological diseases and may also occur in SCA.

The aim of this study is to determine the average dental arch dimension in Ghanaian patients with SCA and compare the findings with those from haematologically healthy individuals.

Methods: The study involved 120 participants comprising 60 individuals with SCA and 60 age and sex-matched haematologically healthy individuals without SCA (controls). Each consisted of 30 males and 30 females aged between 18 and 35 years with a mean age of 25.29 ± 5.32 years. Measurements of dental arch dimensions were made directly on the dental study casts using a digital sliding caliper. Differences in dental arch dimensions between patients with SCA and controls were determined using the Two sample t-test for unpaired data. The level of significance was set at $p < 0.05$.

Results: An assessment of dental arch dimension revealed that the mandibular arch depth of patients with SCA (25.18 ± 2.17 mm) was significantly smaller than those in the controls (25.92 ± 1.74 mm) ($p = 0.04$). Additionally, the mandibular arch length of patients with SCA (69.13 ± 4.89 mm) was reduced compared to controls (70.90 ± 4.19 mm) ($p = 0.03$). Individuals with SCA generally demonstrated smaller means for other parameters assessed, but these differences were not statistically significant.

Conclusion: This study demonstrated the differences in arch diameter parameters between patients with SCA and the haematologically healthy controls and this must be taken into consideration during orthodontic diagnosis and treatment planning for affected individuals.

Keywords: Dental- arch dimensions, Gnathopathy, Sick cell anaemia,

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Introduction

Sickle cell disease (SCD) is a hereditary blood disorder characterized by abnormally shaped red blood cells. Affected individuals usually present with chronic anaemia. The anaemia associated with SCD is caused by an abnormal haemoglobin mutation, haemoglobin (Hb) S, which results in glutamic acid being replaced by valine on the beta chain. This mutation causes chronic haemolytic anaemia and vaso-occlusive episodes with ischemic injury of many tissues.¹ There are several forms of SCD and the principal genotypes include homozygous sickle cell

anaemia (SCA), sickle-haemoglobin C disease, and sickle cell- β -thalassaemia. About 100 million people worldwide are affected by the disease.² There is a particularly high prevalence in Africa and people of African descent around the world.^{3,4} Currently, the prevalence of SCD in Ghana is 2% of all births annually with a carrier rate of 30% of the population.⁵

Clinical manifestations of the disease include both acute and chronic presentations. Acute presentations of the disease include pain crises, acute chest syndrome, priapism and strokes.³ Chronic complications include nephropathies and

chronic bone pain.⁶ SCA has been documented to be associated with craniofacial manifestations amongst which are an increased incidence of osteomyelitis of the jaw bones, bossing of the frontal and parietal bones, and maxillary prognathism (sickle cell gnathopathy).⁷ Oral manifestations of the disease include pulp necrosis, atrophy of the papillae of the tongue, neuropathy of the mental nerve, enamel defects, tooth eruption disturbances, and dental malocclusion⁸.

Malocclusion is a significant dentofacial complication of SCA.⁹ Several studies have demonstrated that malocclusions are more prevalent in individuals with SCA than in the general population.^{7,9,10} Furthermore, individuals with SCA appear to be more prone to developing severe forms of malocclusion than the general population.⁷ Although the aetiology of malocclusion in SCA has not been extensively studied, similar to the development of malocclusion in the general population, there is likely to be a complex interaction of factors which may include skeletal, dental, and soft tissue factors.

Sickle cell gnathopathy (maxillary prominence due to compensatory bone marrow hyperplasia in the maxilla) which occurs in about a third of individuals with SCA¹¹ may have an effect on the arch dimensions of affected individuals. These discrepancies in arch dimensions, in addition to tooth size variations, can contribute to the development of malocclusion in SCA and may result in spacing or crowding of the dentition. Although actual discrepancies in arch dimensions and tooth size have not been studied in detail in SCA, some studies have described a reduction in arch dimensions and mesiodistal tooth width in other haemoglobinopathies.^{12,13} It is therefore plausible that in individuals with Sickle Cell Gnathopathy, arch dimensions may be affected.

Studies on the dentofacial effects of SCA in Ghana are lacking, therefore, this study aim to determine the average dental arch dimensions of Ghanaian patients with SCA and compare the findings with those from unaffected (SCD-free) individuals.

Materials and Methods

Sickle cell disease (SCD) is a hereditary blood disorder characterized by abnormally shaped red blood cells. Affected individuals usually present with chronic anaemia. The anaemia associated with SCD is caused by an abnormal haemoglobin mutation, haemoglobin (Hb) S, which results in glutamic acid being replaced by valine on the beta chain. This mutation causes chronic haemolytic anaemia and vaso-occlusive episodes with ischemic injury of many tissues.¹ There are several forms of SCD and the principal genotypes include homozygous sickle cell anaemia (SCA), sickle-haemoglobin C disease, and sickle cell- β -thalassaemia. About 100 million people worldwide are affected by the disease.² There is a particularly high prevalence in Africa and people of African descent around the world.^{3,4} Currently, the prevalence of SCD in Ghana is 2% of all births annually with a carrier rate of 30% of the population.⁵ Clinical manifestations of the disease include both acute and chronic presentations. Acute presentations of the disease include pain crises, acute chest syndrome, priapism and strokes.³ Chronic complications include nephropathies and chronic bone pain.⁶ SCA has been documented to be associated with craniofacial manifestations amongst which are an increased incidence of osteomyelitis of the jaw bones, bossing of the frontal and parietal bones, and maxillary prognathism (sickle cell gnathopathy).⁷ Oral manifestations of the disease include pulp necrosis, atrophy of the papillae of the tongue, neuropathy of the mental nerve, enamel defects, tooth eruption disturbances, and dental malocclusion⁸.

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Figure 1: Measurement of arch widths.

ICW – Inter-canine width, IPW – Inter-premolar width, IMW – Inter-molar width

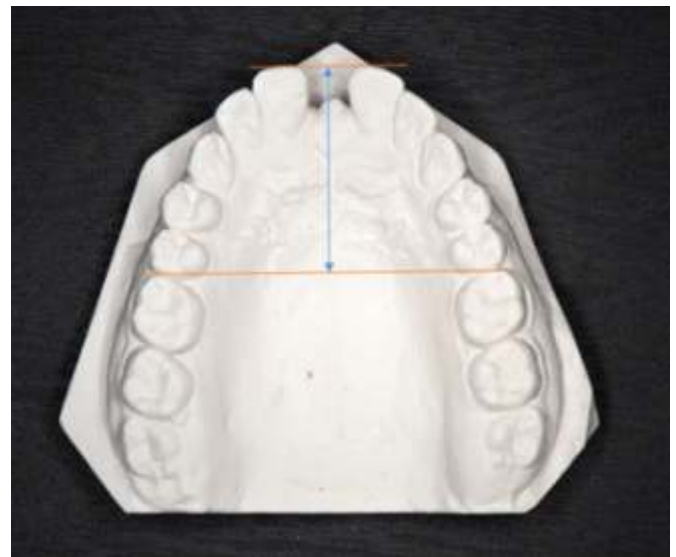


Figure 2: Measurement of Arch depth



Figure 3: Measurement of Arch length



Figure 4: Measurement of palatal height

Assessment of Random Error

The assessment of random error associated with measurements in this study was carried out by repeating all measurements on 25 randomly selected casts after a period of 2 weeks from the initial measurement. The random error was then calculated using Dahlberg's formula¹⁵:

$$D = \sqrt{\sum_{i=1}^N \frac{d_i^2}{2N}}$$

Where D is the Dahlberg error, (d) is the difference between two readings of the same variable and (N) is the sample size that was re-measured.

Table 1 shows the random error for measurements of the maxillary and mandibular arch dimensions. Dahlberg's method showed that the mandibular arch length measurement demonstrated the greatest error (0.31) while the maxillary palatal height demonstrated the least error (0.09).

Table 1: Assessment of measurement error of Arch dimensions by Dahlberg's method

Arch dimensions	Maxillary arch	Mandibular arch
Arch length (mm)	0.169	0.310
Arch depth (mm)	0.126	0.093
Inter-canine width (mm)	0.208	0.115
Inter-premolar width (mm)	0.112	0.102
Inter-molar width (mm)	0.106	0.174
Palatal height (mm)	0.090	NA

Descriptive statistics were presented as mean $\pm$ standard deviation. Differences in arch dimensions and tooth sizes between SCA patients and controls were determined using the Student's t-test for unpaired data. All p-values < 0.05 were judged statistically significant. Data was analyzed using SPSS version 24.

Data collected were entered into and analyzed using Statistical Package for the Social Sciences (SPSS) version 24.0. The data was summarized as the mean $\pm$ standard deviation for continuous variables and percentages for categorical variables. Differences in arch dimensions between individuals with SCA and controls were determined using the Student's t-test for unpaired data. All p-values < 0.05 were judged statistically significant

Results

Dental arch dimensions for patients with SCA and controls are shown in Table 2.

A comparison of maxillary arch dimensions between patients with SCA and controls revealed that mean arch length was reduced in patients with SCA (80.00 ± 5.17 mm) compared to controls (81.38 ± 4.69 mm). Arch depth was also reduced in patients with SCA (29.56 ± 2.66 mm) when compared to the mean value for controls (30.35 ± 2.38 mm). A reduction in the mean inter-canine width was observed in patients with SCA (36.87 ± 3.27 mm) when compared to haematologically - healthy participants (37.42 ± 2.48 mm). The differences in dimensions listed above were, however, not statistically significant. Patients with SCA, on the

other hand, showed increases in the maxillary inter-molar width and palatal height when compared with controls. These differences were also not statistically significant.

When the mean arch dimensions mandibular arch were assessed and compared between individuals with SCA and non-SCD controls it was observed that with the exception of the inter-molar width, where individuals with SCA had average dimensions greater than that of controls, all other arch

dimensions assessed in the mandible showed mean values lower in patients with SCA than in controls. The mean mandibular arch length of patients with SCA was 69.13 ± 4.89 mm, a value that was determined to be significantly lower than that of controls (70.90 ± 4.19 mm; $p = 0.03$). A statistically significant difference in the arch depth was also recorded, with patients with SCA having a reduced average arch depth of 25.18 ± 2.17 mm and non-SCD controls, a value of 25.92 ± 1.74 mm; $p = 0.04$.

Table 2: Arch dimensions of patients with SCA and control participants.

Arch dimension	SCA (mean $\pm$ sd)	Control (mean $\pm$ sd)	P-value
Mx arch length (mm)	80.00 $\pm$ 5.17	81.38 $\pm$ 4.69	0.12
Mx arch depth (mm)	29.56 $\pm$ 2.66	30.35 $\pm$ 2.38	0.08
Mx inter-canine width (mm)	36.87 $\pm$ 3.27	37.42 $\pm$ 2.48	0.30
Mx inter-premolar width (mm)	50.89 $\pm$ 3.28	51.02 $\pm$ 3.15	0.81
Mx inter-molar width (mm)	55.90 $\pm$ 3.31	55.27 $\pm$ 3.14	0.28
Mx Palatal height (mm)	17.88 $\pm$ 2.07	17.19 $\pm$ 2.33	0.08
Mn arch length (mm)	69.13 $\pm$ 4.89	70.90 $\pm$ 4.19	0.03*
Mn arch depth (mm)	25.18 $\pm$ 2.17	25.92 $\pm$ 1.74	0.04*
Mn inter-canine width (mm)	28.33 $\pm$ 3.28	28.71 $\pm$ 2.54	0.47
Mn inter-premolar width (mm)	42.32 $\pm$ 0.54	42.68 $\pm$ 3.21	0.57
Mn inter-molar width (mm)	48.08 $\pm$ 3.41	47.60 $\pm$ 2.89	0.41

Data presented as mean $\pm$ standard deviation *Significance at $p < 0.05$

Mx: maxillary; Mn: mandibular

Discussion

Dental arch dimensions, as demonstrated earlier, exhibit a strong genetic influence. It has however been shown that environmental and pathological factors, among others, can influence the size of the maxillary and mandibular arches¹⁶. Studies exploring

the effect of SCD on arch dimensions were not found in the literature; however, research exploring the effects of haemoglobinopathies such as thalassaemia major have produced interesting results. Thalassaemia major is the most severe form of Beta thalassaemia, a haematological condition that results

in the reduced production of beta-haemoglobin chains. It is usually common in Mediterranean populations and causes severe disease with symptoms similar to SCA.

Hattab *et al.*¹² Kumar *et al.*¹³ and Al-Wahadni¹⁷, demonstrated that in general, individuals with thalassaemia major had reduced dental arch dimensions when compared to haematologically healthy controls.

In this present study, an assessment of maxillary and mandibular arch dimensions revealed that average arch dimensions were generally reduced in patients with SCA compared to controls. With the exception of maxillary and mandibular inter-molar width, as well as the palatal height, all other parameters showed mean values that were reduced in patients with SCA in comparison with controls. These differences were however not statistically significant. Statistically significant differences were however observed for mandibular arch length and arch depth, with patients with SCA exhibiting reduced mean values, compared to the controls. These findings agree with values obtained by Hattab *et al.*¹² who recorded reduced values for arch depth and perimeter in patients with thalassaemia major. Hattab *et al.*¹², however, noticed a smaller difference in mandibular arch depth between cases and controls than in the maxilla where a greater difference in means was observed. This was attributed to the thicker cortical bone in the mandible, which prevented expansion. This finding, however, is contrary to this current study as no significant difference in the maxillary arch depth was recorded between cases and controls.

In this study, it was observed that the mean value for maxillary and mandibular inter-molar widths was greater in individuals with SCA when compared to the controls, although this difference was not statistically significant. These values are at variance with those obtained by Hattab *et al.*¹², Al-Wahadni *et al.*¹³ and Kumar *et al.*¹⁷, who noticed greater values for

inter-molar width in controls when compared to patients with thalassaemia major. These findings suggest that the arch forms of individuals with SCA in this study may be wider posteriorly and narrower anteriorly. An analysis of arch forms was, however, not performed in this study and may constitute a basis for further research.

Additionally, mean palatal height was increased in individuals with SCA when compared to the controls, a difference that was not statistically significant. This finding may, however, suggest an increased vertical development of the maxillary alveolar arches. It is possible that the hyperplasia of the bone marrow thought to cause maxillary prominence in sickle cell gnathopathy may actually result in the vertical development of the maxillary alveolar bone and cause an increase in measured palatal height. Further studies exploring this hypothesis would however be required.

The findings of our study also appear to conflict with studies that have suggested an increased prominence of the maxilla with proclination of teeth and spacing in individuals with SCA.¹¹ Those findings would logically have translated to a general increase in arch dimensions in patients with SCA compared to the controls, however, the converse was observed to be the case in this study. Some cephalometric studies have suggested that the increase in maxillary prominence observed in SCA may be due to a reduced cranial base length associated with a retrognathic mandible and not maxillary prognathism as initially thought.¹⁸ A possible cause for reduced mandibular arch depth and length in patients with SCA compared to controls could be the growth retardation that occurs in SCA. Studies have demonstrated that individuals with SCA generally grow and develop more slowly, often reaching puberty later than their peers. Additionally, adults

with SCA are generally shorter and thinner than the general population.^{19,20} The reduced mandibular arch length and depth in this study could therefore be a reflection of general growth retardation that occurs in SCA.

Conclusion

In this study, a comparison of average arch dimensions between individuals with SCA and controls without SCD revealed generally reduced mean values for individuals with SCA compared to the controls with statistically significant differences noted for mandibular arch length and depth. It is therefore recommended that proper orthodontic diagnosis and treatment planning be performed, especially in patients with SCA, as their arch dimensions may not match that of the general population.

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Acknowledgements

We wish to thank all the participants who took part in this study. We would also like to acknowledge the nurses and resident doctors of the Orthodontic and Paedodontics department of University of Ghana Dental School for their assistance with data collection.

Contributions

Contributions were made by all authors to data collection, analysis, and write-up of this study. The final manuscript was assessed and approved by all authors.

Funding/grants

This study was funded solely by the authors.

Conflict of interest

None declared.

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