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Radiological Assessment of Craniometric Indices in Sickle Cell Anaemia Subjects in Rivers State.

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ABSTRACT

This study was carried out with the aim of radiologically determining any differences in the cephalometric and facial indices between SCA and non-SCA subjects of same age and sex and determine values for cephalic and Nasal Indices of SCA subjects. A total of 286 skull radiographs of subjects aged 3 – 30 years, comprising 250 radiographs of non-SCA and 36 radiographs of SCA patients were used for this study. Measurements were carried out using a Digital Venier's Caliper to determine values for calculation of cephalic and Nasal indices for the different ages and sexes. Occipito-frontal and Lateral radiographs of non-SCA subjects were obtained from the Radiology Departments of the University of Port Harcourt Teaching Hospital (UPTH) and Braithwaite Memorial Specialist Hospital (BMSH) while radiographs of SCA subjects were shot at the Radiology Department of UPTH. Occipito-frontal and Lateral radiographs of non-SCA subjects were obtained from the Radiology Departments of the University of Port Harcourt Teaching Hospital (UPTH) and Braithwaite Memorial Specialist Hospital (BMSH) while radiographs of SCA subjects were shot at the Radiology Department of UPTH. The data were statistically analyzed using Z-test at 95% confidence interval. From the results obtained, mean cephalic index of non-SCA subjects was found to be 76.66±10.67 while those of SCA subjects was 76.21±3.11 which was not statistically significant($p>0.05$). Male mean cephalic index for non-SCA subjects was determined as 78.29±4.72 while SCA subjects was 75.99±3.22 and was statistically significant($p<0.05$). Female non-SCA subjects mean cephalic index was found to be 76.91±6.46 while that of SCA subject was 76.40±1.14 (not significant, $p>0.05$). Mean Nasal index of non-SCA subjects was 74.71±9.23 and SCA subjects was 72.90±4.80. The results indicated growth retardation in SCA subjects which can be used as diagnostic tool in the management of sickle cell anaemia patients by maxillofacial surgeons, paediatricians, medical radiologists and forensic anthropologists.

Key words: Cephalic and Nasal Indices, Radiology, Rivers State, Sickle Cell Anaemia

INTRODUCTION

Sickle cell anaemia is characterized by the unusually "sickle" shaped red blood cells found in the blood stream. Life expectancy is shortened, with studies reporting an average life expectancy of 42 in males and 48 in females[1]. Interest in skull and bone lesions associated with severe anaemia was stimulated in 1925 when Cooley and Lee described striking radiographic abnormalities in the bones of children with the disease-thalassemia major. Observation of some lesions in the skull of sickle cell anaemic patients were also reported[2]. A monograph by Reynolds[3] reveals a noteworthy coverage of the radiographic features of sickle cell anaemia and related haemoglobinopathies. Anatomic lesions in the skull and other bones have been reported[4].

Anthropometric deficits have been indicated on craniofacial structures (prognathic maxillary profile, frontal bossing, depression in bridge of nose, malocclusion of teeth, etc) of sickle cell disease subjects in Nigeria[5,6] and other parts of the world[7]. It has also been indicated that sternomental distance of sickle cell anaemia subjects is significantly reduced when compared with the non-sickle cell anaemia subjects[8]. A non radiological study on school children in Benin City, Nigeria, reported the cephalic index as 80.86. Sexual dimorphism was observed as male cephalic index value (81.15) which was higher than female cephalic index value (80.58) as reported by the study [9].

The aim of the study was to determine whether there are any differences in the cephalometric and facial indices between Sickle Cell Anaemia (SCA) and non – sickle cell anaemia subjects of same age and sex. It was also aimed at determining, radiologically, possible cephalic, Nasal indices range of values for sickle cell anaemia subjects in Rivers State, Nigeria.

MATERIALS AND METHODS

This study was carried out in Port Harcourt, Rivers State, Nigeria, between November, 2010 and April, 2011. A total of 250 radiographs of non-sickle cell

anaemia subjects were obtained from University of Port Harcourt Teaching Hospital (UPTH) and Braithwaith Memorial Specialist Hospital (BMSH). Radiographs of the skull of 36 sickle cell anaemia patients aged 3 – 30 years were shot in radiology department of the University of Port Harcourt Teaching Hospital. Each skull radiograph obtained (Anteroposterior and Lateral projections as described by Stewart *et al.,*) [10] was placed on a lighted x-ray viewing box to make clear the markings on the skull using a sharp pencil. A digital vernier caliper was adjusted and placed on the radiograph to measure the required parameters in mm (demonstrated in photo plates I -VI) as outlined below using the method by Del Sol for Cephalic Index [11]; Romo and Abraham for Nasal Index [12].

The **Cephalic Index** (A1/B1×100) was calculated from parameters [head breadth from euryon (eu) to euryon (eu), and head length from glabella (g) to opistocranion (op)] measured using a Digital Vernier Spreading Caliper as shown in photo-plates I and II .

The **Nasal Index** (A4/B2×100) was calculated from parameters measured using a Digital Vernier Caliper (photo-plates III and IV). Nasal length was taken as the distance from nasion to the nasospinale while the nasal breadth was the distance from ala (al) to ala (al) [12].

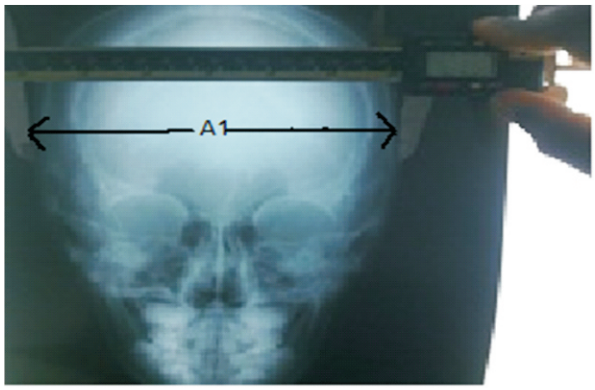

Photo-Plate I: Measurement of Head Breadth

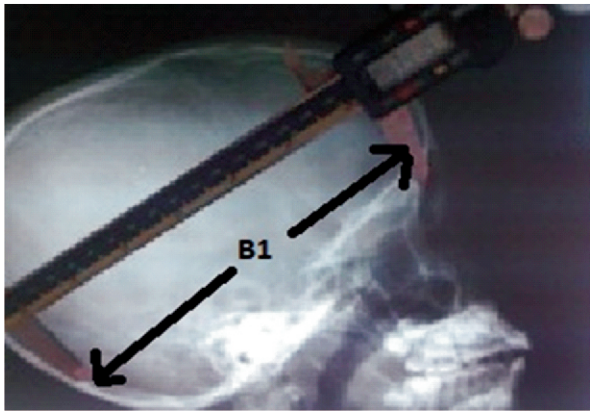

Photo-Plate II: Measurement of Head Length

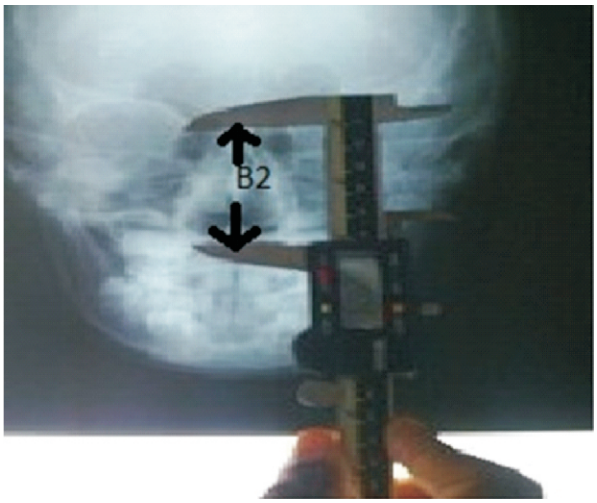

Photo-Plate III: Measurement of Nasal Height

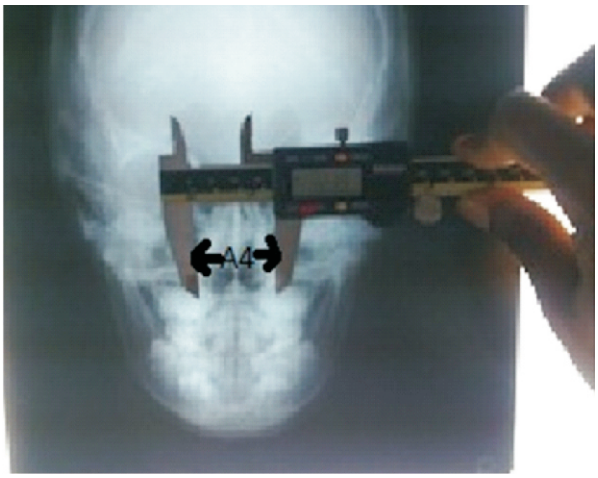

Photo-Plate IV: Measurement of Nasal Breadth

Ethical Clearance: Ethical clearance was sort and obtained from the Ethics Committee of the University of Port Harcourt Teaching Hospital and Informed Consent obtained from Parents of Sickle Cell Anaemia subjects.

Data Analysis

The data was analysized with descriptive statistics: mean, standard deviation. The Z-Test was used for test of significance of the various parameters described above. The data obtained was analyzed for central tendency for these craniofacial measurements and their measure of dispersion from the Mean determined.

The sample size was determined using the formula for minimum sample size AAPOR [13]:

1.) $N = (Zi\text{-}a)^2 P(1\text{-}P) / d^2$

N= Minimum Sample Size

a= Significant level of effort tolerable for this study at 0.05 confidence level

$Zi - a = 1.96$

P= Best estimate of prevalence from

literature review set at 0.02

d= Degree of accuracy desired set at 0.05

$N = 1.96^2 \times 0.02(1-0.02) / 0.05^2$

$N = 30.12$

2.) The Standard Deviation Formular

$$S = \sqrt{\frac{\sum_{i=1}^{N} (x_i - \bar{x})^2}{N-1}}$$

3.) Z(calculated)

$$= \frac{(\bar{x}_1 - \bar{x}_2)}{\sqrt{\frac{\sigma_1^2}{n1} + \frac{\sigma_2^2}{n2}}}$$

where σ = standard deviation of sample size

RESULTS

All data were analyzed statistically and separately and the Mean and Standard Deviation obtained for the various parameters for the different age groups and sex for the sickle cell anaemia and non-sickle cell anaemia growing children summarized in tabular and graphical forms. The results were subjected to test of significance using the Z-test hypothesis with variance set at infinity and a 95% confidence interval (P = 0.05). The results obtained were tabulated and analyzed as shown in **Tables 1-2 and Figures 1 and 2 below.** The radiological determination of craniofacial parameters in sickle cell anaemia patients of this study showed mean cephalic index for non-Sickle Cell Anaemia (non-SCA) subjects to be 77.66±10.67 and that of sickle cell anaemia (SCA) subjects as 76.21±3.11. Values obtained for SCA mean cephalic index in all age groups were lower than values obtained for non-SCA as shown on the graph in figure 1. This places both non-SCA and SCA in mesocephalic group of cephalic index. When male non-SCA subjects were compared with male SCA subjects (table 2), there was a generally lower mean cephalic index recorded for male SCA subjects in all age groups measured; with age group 18-22 tending towards dolicocephaly (figure 3); as against a much higher value in females mean cephalic index (figure 4). There was no statistical significant difference ($p>0.05$) in all age group measured for males.

The Mean Nasal Index of non-SCA subjects determined radiologically in this study was 74.71±9.23 and that of SCA subjects was 72.90±4.80. Mean Nasal Index of male non-SCA subjects in all age groups recorded for this study ranges between 70.72 to 80.88 (table 5) while that of the SCA subjects ranges between 70.41 to 73.46 as shown in table 6. It therefore shows a mesorrhine type of nose for both non-SCA and SCA subjects, however, the values for SCA subjects were generally lower in male subjects (table 5) except in age group 13-17 (as clearly indicated on the graph in figure 5) when compared with non-SCA subjects. Similarly, the female SCA subjects showed lower values of Nasal Index (table 6) except in age group 3-7 (clearly indicated on the graph in figure 6). Statistically significant difference was recorded in age group 8-12 for both male and females (tables 5 and 6) with male mean nasal index value for sickle cell anaemia subjects as 73.46±4.94 and table of female as 64.68 ± 4.31.

Table1 : Mean Cephalic Index of Non-SCA and SCA Subjects for Different Age Groups

Age Group	Non-SCA		SCA	
	N	Mean Cephalic Index ±SD	N	Mean Cephalic Index ± SD
3-7	85	77.16 ± 4.48	9	76.66 ± 3.70
8-12	27	78.68 ± 4.56	13	76.39 ± 3.84
13-17	36	79.03 ± 3.97	8	76.05 ± 3.90
18-22	21	77.53 ± 4.74	6	75.75 ± 0.99
23-27	31	75.92 ± 11.56	-	-
28-30	50	77.66 ± 4.67	-	-
TOTAL	**250**		**36**	

N=Sample Size, SD = Standard Deviation, SCA = Sickle Cell Anaemia

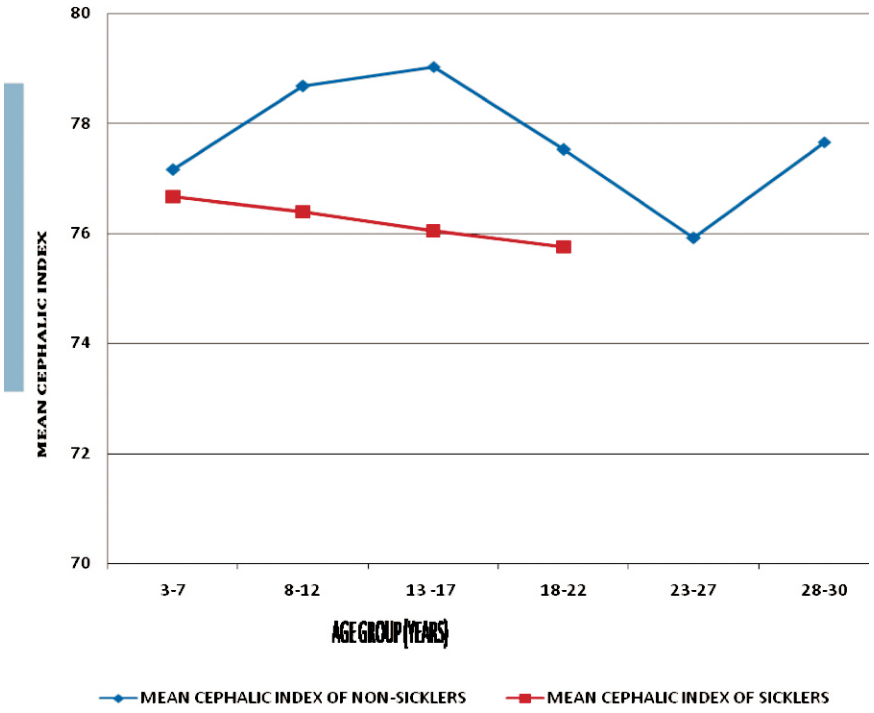

Figure 1: Mean Cephalic Index Against Age Group of Non-SCA and SCA Subjects

Table 2: Mean Nasal Index of Non-SCA and SCA Subjects for Different Age Groups

Age Group	Non- SCA		SCA	
	N	Mean Nasal Index ± SD	N	Mean Nasal Index ± SD
3-7	85	75.37 ± 8.90	9	74.63 ± 7.55
8-12	27	77.44 ± 8.65	13	71.44 ± 6.06
13-17	36	74.92 ± 13.48	8	73.68 ± 4.17
18-22	21	72.69 ± 8.16	6	71.84 ± 1.43
23-27	31	74.89 ± 7.63	-	-
28-30	50	72.93 ± 8.54	-	-

N= Sample Size, SD= Standard Deviation, SCA = Sickle Cell Anaemia

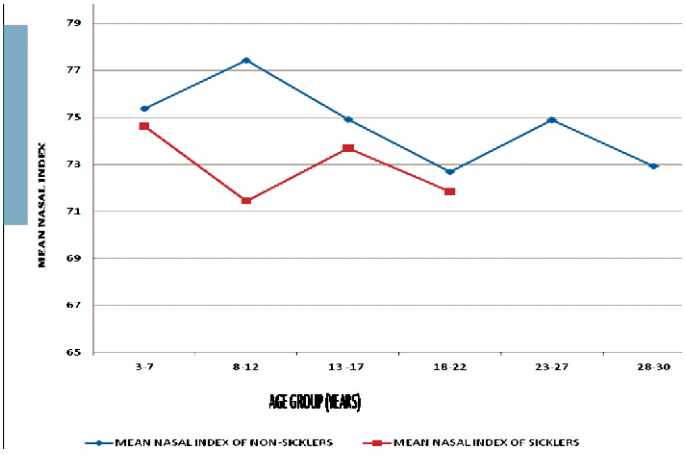

Figure 2 : Mean Nasal Index Against Age Group Of Non-SCA and SCA Subjects

Table 3: Mean Cephalic Index of Non- SCA and SCA Males Subjects for Different Age Groups.

Age Group	Male Non- SCA		Male SCA		Level of Significance
	N	MCI ± SD	N	MCI ± SD	
3-7	51	76.73 ± 4.54	8	76.61 ± 4.24	P > 0.05
8-12	14	79.05 ± 5.52	10	76.22 ± 4.18	P > 0.05
13-17	18	78.80 ± 3.58	5	76.36 ± 4.46	P > 0.05
18-22	11	78.35 ± 4.77	2	74.76 ± 0.40	P > 0.05
23-27	21	78.27 ± 5.37	-	-	-
28-30	34	78.57 ± 4.56	-	-	-

N = Sample Size, SD = Standard Deviation, MCI = Mean Cephalic Index, SCA = Sickle Cell Anaemia

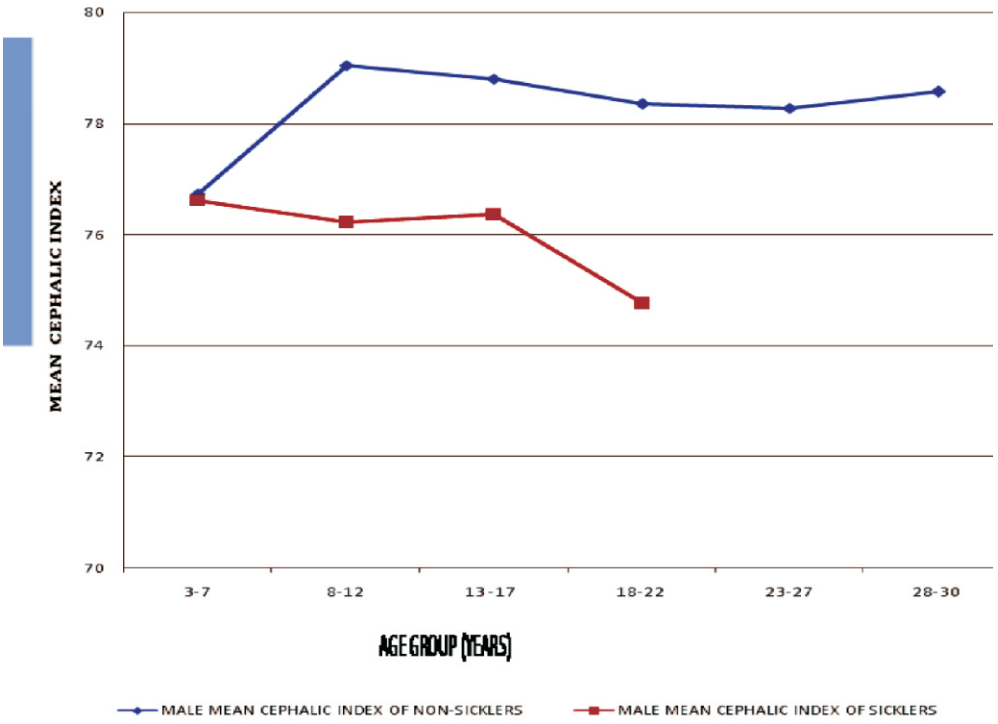

Figure 3: Mean Cephalic Index Against Age Group Of Male Non- Sca And Male Sca Subjects

Table 4: Mean cephalic Index of Non- SCA and SCA Females Subjects for Different Age Groups

Age Group	Female Non- SCA		Female SCA		Level of Significance
	N	MCI ± SD	N	MCI ± SD	
3-7	34	77.79 ± 4.29	5	76.76 ± 2.25	P > 0.05
8-12	13	78.28 ± 3.17	3	76.97 ± 2.32	P > 0.05
13-17	18	79.23 ± 4.38*	2	76.57 ± 1.45*	P < 0.05*
18-22	10	76.63 ± 4.53	1	76.74 ± 0.00	P > 0.05
23-27	10	70.98 ± 17.83	-	-	-
28-30	34	78.57 ± 4.56			

N = Sample Size, SD = Standard Deviation, * Statistically Significant, SCA = Sickle Cell Anaemia

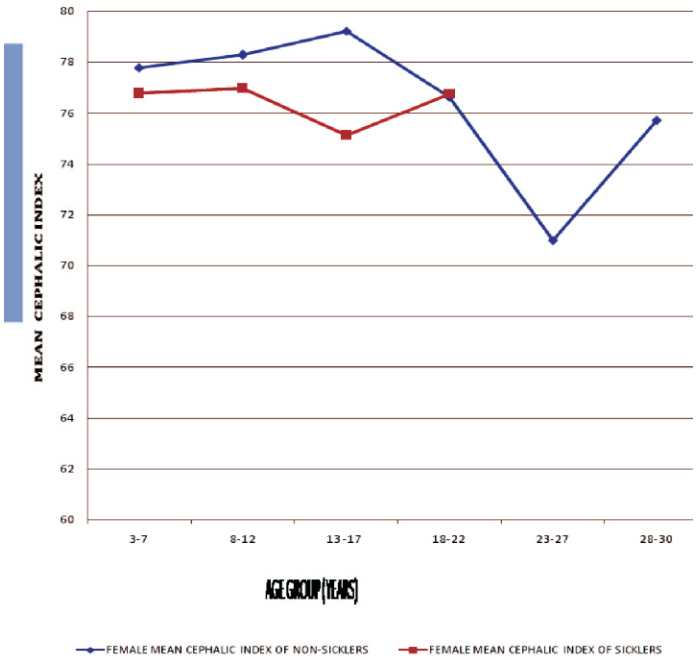

Figure 4: Mean Cephalic Index Against Age Group Of Female Non- Sca And Female Sca Subjects

Table 5: Mean Nasal Index of Non- SCA and SCA Males Subjects for Different Age Groups

Age Group	Male Non- SCA		Male SCA		Level of Significance
	N	Mean Nasal Index ± SD	N	Mean Nasal Index ± SD	
3-7	51	75.67 ± 8.46	8	72.20 ± 8.15	P > 0.05
8-12	14	80.88 ± 8.10*	10	73.46 ± 4.94*	P < 0.05*
13-17	18	71.61 ± 12.76	5	73.14 ± 4.69	P > 0.05
18-22	11	70.72 ± 7.59	2	70.41 ± 0.40	P > 0.05
23-27	21	73.95 ± 7.17	-	-	-
28-30	34	75.34 ± 7.71	-	-	-

N = Sample Size, SD = Standard Deviation, * Statistically Significant, SCA = Sickle Cell Anaemia

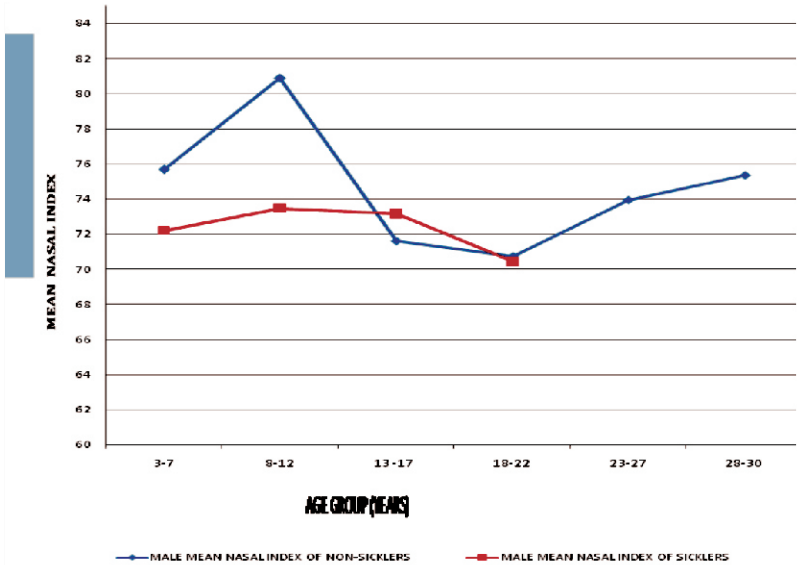

Figure 5: Mean Nasal Index Against Age Group of Male Non- SCA and Male SCA Subjects

Table 6: Mean Nasal Index of Non- SCA and SCA Females Subjects for Different Age Groups.

Age Group	Female Non- SCA		Female SCA		Level of Significance
	N	Mean Nasal Index ± SD	N	Mean Nasal Index ± SD	
3-7	34	74.94 ± 9.50*	5	79.49 ± 1.61*	P < 0.05*
8-12	13	73.73 ± 7.63*	3	64.68 ± 4.31*	P < 0.05*
13-17	18	78.22 ± 13.36	2	74.08 ± 1.24	P > 0.05
18-22	10	74.85 ± 8.22	1	73.27 ± 0.00	P > 0.05
23-27	10	76.86 ± 8.16	-	-	-
28-30	16	67.80 ± 7.95	-	-	-

N = Sample Size, SD = Standard Deviation, * Statistically Significant, SCA = Sickle Cell Anaemia

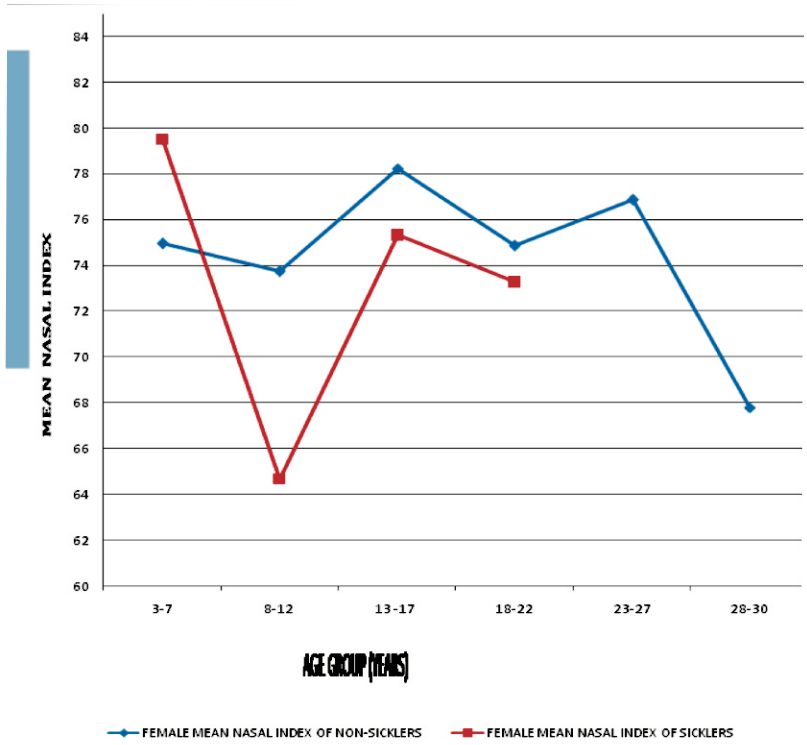

Figure 6: Mean Nasal Index Against Age Group of Female Non- SCA and Female SCA Subjects

DISCUSSION

Development of the cranium and facial structures are known to be affected by sickle cell anaemia from studies carried out, as reported previously [2,4]. Sickle cell anaemia causes a wide range of abnormalities in the skull, especially the calvaria, such as diploic expansion, external or outer table thinning, vertical trabecular striations. Factors that may be responsible include cellular hyperplasia or circulatory factors [14].

It should be noted that diploic expansion of the skull bones progresses with increase in age and occurs mostly in the parietal bone followed by frontal bone[14]. This therefore explains the progressive decrease in the mean cephalic index values measured in sickle cell anaemia subjects (as against the non-sickle cell anaemia subject) seen in tables 3 and 4 as the age group increases in this study. In female comparison of mean cephalic index (table 4), there was a generally lower values recorded for SCA subjects with statistical significant difference ($p<0.05$) observed in age group 13-17 (table 4). This finding agrees with the findings of Fawehinmi and Ligha[9] except in age groups 3-7 and 8-12. The measured values from radiographs of non-SCA and SCA subjects showed a slight variation but still maintain a consistent trend of mesocephaly. The possible explanation for this is the significant diploic thickening in frontal and parietal region of the skull which does not involve squamous portion of temporal or occipital bone [15], giving the SCA subjects lower cephalic index recorded in this study.

Moreover, growth deficit tends to be greater in width or breadth than in height or length of skull and is more

severe in patients with sickle cell anaemia and S-β0 thalassemia than in those with HbSC disease and Sβ thalassemia [15]. This could have also probably accounted for the lower cephalic index found amongst the sickle cell disease patients. Although, in non radiological measurement of mean cephalic indices of various tribes, there are gradual shift toward brachycephalization [16,17], in this study however, mesocephaly is more demonstrated in all age groups measured. Many factors may be responsible for this observed phenomenon: diploic expansion due to marrow hyperplasia and non radiological measurement of subjects' bone dimensions may be affected by tissue and fascia. The mean nasal index value determined radiologically in sickle cell anaemia subjects was lower than that of the non sickle cell anaemia subjects, which was statistically significant in male subjects. When compared in age groups, males showed statistical significant difference in age group 8 – 12 (table 5) while female showed statistical significant difference in age groups 3 -7 and 8 -12 (table 6). This agrees with findings reported by Fawehinmi and Ligha [6]. Possible reason for this lower values observed could be the depression of bridge of the nose[7] and maxillary overgrowth due to bone marrow hyperplasia which are common facial characteristic developed by sickle cell anaemia patients[5]. This could result to greater nasal width recorded in homozygous sickle cell anaemia patients and may be expected between the age groups 3-7 and 8-12 as evidenced in this study (tables 5 and 6).

Many factors are known to influence the manifestations of sickle cell anaemia, among which are environmental factors like pollution, poor sanitary conditions, personal hygiene and other poor social circumstances[18]. Determinants of disease severity include internal factors such as hyperplasia of bone marrow which may cause osteopenia and growth disturbances[19], external environmental influences, socio-cultural variables such as widespread poverty, poor state of health facilities, illiteracy, ignorance and the African view of disease causation. This is likely to be the reasons why the severity of sickle cell disease is more in the sub-saharan Africa[1]. An understanding of the pathophysiology of sickle cell disease and knowledge of the various clinical and radiologic manifestations are crucial for prompt diagnosis and appropriate treatment [20].

CONCLUSION

The indices of the parameters of the different age groups are generally lower when compared to their non-SCA counterparts, which is an indication that growth rate is slower in homozygous sickle cell anaemia patients.
Data obtained can also be used as diagnostic tools in the management of sickle cell anaemia patients by maxillofacial surgeons, paediatricians, medical radiologists and forensic anthropologists.

RECOMMENDATIONS

1. This comparative study of radiological assessment of craniofacial indices of both SCA and non-SCA subjects should be extended to other parts of Nigeria, Africa and Negroid in other parts of the world.
2. This comparative study should be subjected to further investigations especially other radiological techniques to see how this haemoglobinopathy affects the craniofacial indices in these subjects.

ACKNOWLEDGEMENT

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