Anthropometric findings in Nigerian children with sickle cell disease

Folakemi A. Oredugba, BDS, FWACS, MPH   Kofo O. Savage, BDS, MPH, MSc, FMCDS, FWACS

Dr. Oredugba is lecturer, Department of Child Dental Health; Dr. Savage is senior lecturer,
Department of Preventive Dentistry, College of Medicine, University of Lagos, Lagos, Nigeria.
Correspond with Dr. Oredugba at tored@nova.net.ng

Abstract

Purpose: Sickle cell disease (SCD) subjects have been widely reported to present with various anthropometric deficits, including malocclusion and stunting, compared to their unaffected peers. This study was carried out to examine these reports in Lagos.

Methods: A hospital-based cross-sectional study was carried out in 3 outpatient SCD units of one teaching and two general hospitals. All established SCD subjects aged 1 to 18 years (n=177) were examined on consecutive clinic days. Controls (unaffected subjects; n=122) were obtained from well-baby and surgical emergency clinics. Facial profile, occlusion, height, weight, head and mid-upper arm (MUA) circumference measurements were noted.

Results: A prognathic maxillary profile was found to be more prevalent in SCD subjects (21%) than controls (4%; *P*<.05). Class II malocclusion was found in 21% of SCD subjects compared to 2% of controls (*P*<.05). The overall mean height, weight and head circumference of SCD subjects and controls were not significantly different (*P*>.05). However, at age 18 years, the weight of the SCD group was significantly less than that of the control group (*P*<.05). The mean MUA circumference was significantly higher in the control group (20.04 cm±3.80) than in SCD subjects (17.91 cm±2.96; *P*< .05).

Conclusions: There was a higher prevalence of maxillary protrusion and Class II malocclusion in SCD subjects than controls. Height and weight were not significantly different in both groups except at 18 years when SCD subjects weighed less than controls. (*Pediatr Dent.* 2002;24:321-325)

KEYWORDS: GROWTH, SICKLE CELL DISEASE, NIGERIAN CHILDREN

Sickle cell disease is a generic term for the family of hemoglobin (Hb) disorders having in common the inheritance of the sickle cell b-globin gene (HbS) from at least one parent.[1] Sickle cell anemia (SCA) results from the inheritance of the HbS gene from both parents, resulting in the homozygous state (HbSS).[1]

It is a condition prevalent among the black race. It is common in Africa, the Mediterranean countries and India and in the descendants of people in these countries who have migrated to Europe, Bahrain and South, Central and North America.[2] It is also a common community health problem in tropical Africa,[3] with a prevalence of 2% in Nigeria.[4]

Clinical features

Sickling of red blood cells (RBC) leads to increased mechanical fragility and reduced red cell survival time. The affected RBCs are thus prone to hemolysis and phagocytosis by the reticuloendothelial system (RES). Consequently, there is hemolytic anemia and jaundice.

This pathophysiology presents some acute and chronic problems, referred to as sickle cell crisis. Acute problems include painful vasoocclusive crisis of bones and joints, especially long bones, metacarpals and metatarsals with overlying soft tissue involvement (the hand-foot syndrome), sequestration crisis, aplastic crisis and hyperhaemolytic crisis.[5] The bone marrow responds to the aplastic crisis by a compensatory hyperplasia.

Gnathopathy, skull bossing, finger clubbing and lymphadenopathy were common presentations in Africans with SCD.[6] The gnathopathy has been found to be progressive and common in those patients with the most severe chronic hemolysis. It is evidenced by maxillary prominence and

Table 1. Lip Seal Pattern of Subjects in the Study*

	SCD	%	Control	%	Total	%
+	97	83	120	98	217	91
-	20	17	2	2	22	9
Total	117	100	122	100	239	100

+=Where the lips are competent
- =Where the lips are not competent
*Chi-square 17.0; P<.05

Table 2. Facial Profile of Subjects in the Study*

Profile	SCD	%	Control	%	Total	%
Normal	93	79	114	93	207	87
Maxillary prognathism	24	21	5	4	29	12
Mandibular prognathism	0	0	3	3	3	1
Total	117	100	122	100	239	100

*Chi-square 17.48; P<.05

Table 3. Occlusion Pattern of Study Population*

Occlusion	SCD	%	Control	%	Total	%
1	90	77	115	94	205	86
2	25	21	3	2	28	12
3	2	2	4	3	6	2
Total	117	100	122	100	239	100

*Chi-square 26.27; P<.05

malocclusion and has been described as the main jaw abnormality seen in SCD patients. Altemus and Elps[7] also reported a tendency towards mandibular retrusion and maxillary protrusion in SCD patients. It was also reported in another study that there is a larger angle of convexity–a measure of protrusion of the maxillary part of the face–to the total profile in sickle cell patients than in controls.[8] The incisors were also seen to be more retruded due to increased lip pressure. Some authors found the maxillary protrusion to be due to an increase in the mean PAR (palate-alveolar ridge) angle.[9] However, it has been suggested that the gnathopathy is most likely a combination of both maxillary prominence and a degree of mandibular retrognathism.[10]

There are conflicting reports in the few reports available on the height and weight of children with SCD. It is reported that they are born with a normal birth weight and length and that anthropometric differences between SCD patients and controls only emerge with age.[1] In a study on the natural history of SCD, the bone age of the patients was found to be delayed between 12 and 19.5 years.[11] They have also been reported to show an average reduction in weight, height, sitting height, limb length, interacromial and intercristal diameter, and skin-fold thickness.[12] Several other studies have also reported a reduction in their weight and height compared with controls.[13-18] Chronic illness, hyperdynamic blood circulation and other ill-defined processes were suggested as contributory.[12] These factors were reported to be operative by age 4 to 6 years. The growth stunt was also reported to be due to previous infarction of the growing ends of bones before the epiphyses unite.[19]

The purpose of this study is to assess these health parameters in subjects with SCD in Lagos, Nigeria, and to compare them with control subjects in the same location, to provide useful information in the provision of adequate health care for SCD patients.

Methods

Sickle cell disease patients, already diagnosed by electrophoresis, attending the SCD clinics at the Lagos University Teaching Hospital, General Hospital, Lagos, and Massey Street Children Hospital, Lagos, were seen on consecutive clinic days. They were all Nigerians aged 1 to 18 years. SCD patients in crisis were excluded from the study. Control subjects (the unaffected) aged 1 to 18 years were seen in the surgical outpatient clinics and well baby clinics of the hospitals. Those without genotype records were excluded from the study. Efforts were made to match for gender and socioeconomic class. Assessments were carried out by one of the authors (FAO).

Information regarding age, gender and parent's occupation and education was obtained. Socioeconomic class was determined by classifying the educational level of each child's mother or father (if the child does not live with his or her mother) into:

- Upper class–code 1: those who attended tertiary institutions.
- Middle class–code 2: those who attended high school.
- Lower class–code 3: those who attended only primary schools or none.[20]

Occupation was not used as an index of socioeconomic class because of the high rate of unemployment in the country. People generally do anything to earn income.

Parameters assessed include facial profile, lip competence, occlusion, overbite, overjet, weight, height, head and

Table 4. Overbite and Overjet in the Study Population

	Overbite*				Overjet†			
	SCD	%	Control	%	SCD	%	Control	%
< 2.5 mm	82	70	113	93	78	67	111	91
2.5 – 3.5 mm	30	26	9	7	33	28	10	8
>3.5 mm	5	4	0	0	6	5	1	1
Total	117	100	122	100	117	100	122	100

*Chi-square 21.13; P<.05
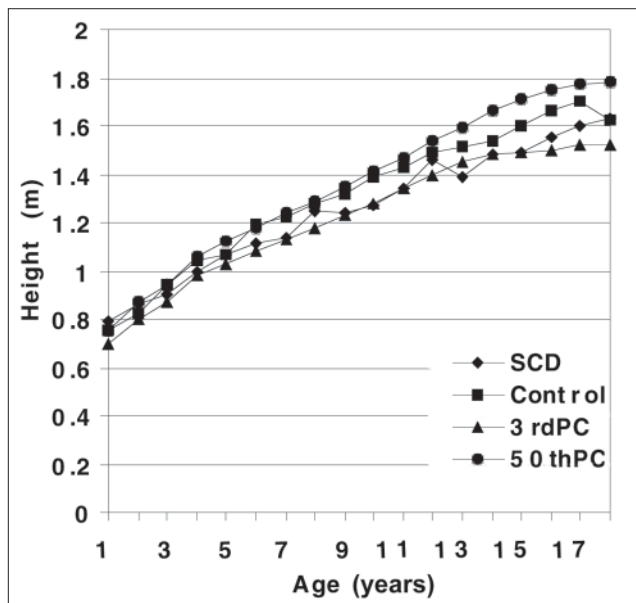†Chi-square 21.51; P<.05

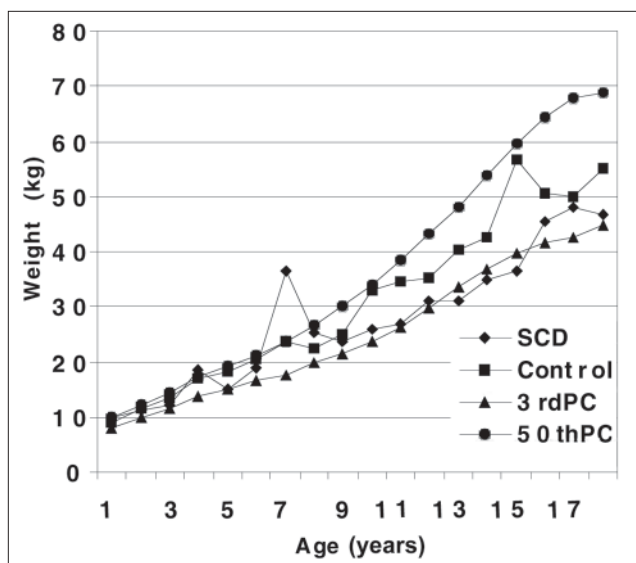Fig 1. Height (m) of subjects compared with anthropometric reference standard

Fig 2. Weight (kg) of subjects compared with anthropometric reference standard

mid-upper arm (MUA) circumference. The facial profile was assessed with each patient relaxed and sitting upright looking at a distant object.[21] The relationship between two lines was viewed: one line dropping from the bridge of the nose to the base of the upper lip; the other line extending from that point downward to the chin. When these line segments form a straight line, it is described as normal. An angle between them indicates either profile convexity (maxillary prognathism) or concavity (mandibular prognathism).

Evaluation of lip competence was carried out with the patient's lips relaxed. If the patient strained to bring the lips together, it was recorded as lip incompetence.[21]

Oral examinations were carried out in daylight using sterile mouth mirrors, explorers and tweezers. One Hanson, UK Scale and a meter rule were used for the weight and height measurements respectively.

Data obtained were analyzed using the EPI Info Version 6.[22] Comparisons were made between affected subjects and controls and socioeconomic class where appropriate. Weight and height were compared using the Anthropometric Reference Standard[23] (ARS). Observed differences were subjected to the chi-square test, ANOVA and Student t test at $P<.05$ level of significance.

Results

A total of 239 subjects, comprising 117 SCD subjects and 122 controls, were examined. Mean age was 10±4.7 years. Females represented a slightly higher percentage of the study population (55%). The majority of the subjects were from the lower SEC (72%).

Lip incompetence was found in 17% of SCD subjects compared with 2% of controls (Table 1). A higher proportion of controls (93%) had a normal facial profile compared with SCD subjects (79%). The SCD subjects showed a higher prevalence of maxillary prognathism (21%) than controls (4%; $P<.05$; Table 2).

Class II malocclusion was found to be more prevalent in the SCD group (21%) than controls (2%; Table 3). Over 25% of SCD subjects also showed increased overbite and overjet values compared with less than 10% of controls (Table 4).

When compared with the ARS, the height and weight values in SCD subjects were near the third percentile while those of controls were near the 50th percentile (Figs 1 and 2). However, there was no statistically significant difference in the overall height and weight values between the two groups ($P>.05$). At age groups of <6 years and 12 years, height and weight were not significantly different. At age 18 years, the mean weight of SCD children was lower than that of controls ($P<.05$; Table 5). There was no significant difference in the mean head circumference of both groups ($P>.05$). The MUA circumference was significantly less in SCD subjects (17.9 cm) than in controls (20.0 cm; $P<.05$). When compared across socioeconomic class, only the head circumference was significantly reduced in the lower SEC of the SCD group (ANOVA; $P<.05$; Table 6). Adequate comparison across the SEC could not be done because few people from the higher SEC make use of these government-owned health care facilities.

Discussion

In this environment, SCD children are assumed to have a characteristic look—a lack of lip seal, which exposes the anterior teeth, and a prognathic maxilla. Findings from this study confirm this speculation. A considerable proportion of the SCD subjects in this study presented with incompetent lips compared to controls. Little has been found in the literature about their lip pattern. This parameter often reflects facial profile and occlusion.

Table 5. Mean Height, Weight and Age of Subjects in the Study				
Age(yrs)	Mean height(m)	P	Mean weight(kg)	P
<6				
SCD	0.94±0.10		13.80±3.43	
Control	0.98±0.12	>.05	15.60±3.62	>.05
12				
SCD	1.46±0.27		31.00±11.31	
Control	1.50±0.11	>.05	35.28±7.41	>.05
18				
SCD	1.64±0.07		47.18±4.89	
Control	1.62±0.08	>.05	55.14±8.03	<.05*

*Statistically significant

Table 6. Anthropometric Values and Socioeconomic Class (SEC)		SCD		Control	
	SEC	Mean	P	Mean	P
Height(m)	1	1.09±0.00		1.33±0.25	
	2	1.34±0.25		1.38±0.30	
	3	1.30±0.25	>.05	1.29±0.23	>.05
Weight(kg)	1	19.00±0.00		33.12±15.21	
	2	32.44±15.74		35.60±14.93	
	3	28.51±12.61	>.05	30.07±13.78	>.05
Head circumf-erence(cm)	1	51.00±0.00		52.34±2.25	
	2	53.47±3.03		53.18±2.41	
	3	51.88±2.34	<.05*	52.18±2.44	>.05
Mid-upper arm circumf-erence (cm)	1	15.00±0.00		20.90±4.16	
	2	18.82±3.57		20.56±3.24	
	3	17.79±2.83	>.05	19.62±3.94	>.05

*Statistically significant

A significant proportion of the SCD group exhibited maxillary prognathism compared with the control group, which supports earlier studies.[6-10] However, the profile measurement in this study could not link this to mandibular retrusion. Overbite and overjet values were significantly increased in the SCD group. This reflects incisor protrusion, in contrast to earlier studies.[8] Lip pressure might become a consideration as the patient grows older and becomes more conscious of the anomaly.

Expectedly, Class II malocclusion was prevalent in the SCD subjects because of the maxillary protrusion and increased overbite and overjet. Few studies have been done on the occlusion in SCD patients.

In Nigeria, affected children as well as adults are believed to be of lower than average body weight and height than their unaffected peers.[17,18] The result from this study shows the same trend in height and weight pattern in both groups in the first five years. After this period, there appears to be growth differences. Such growth deficits in sickle cell patients have been reported by earlier studies to be more pronounced with increasing age.[18] This report is supported by the significantly reduced weight of the SCD group at age 18 years ($P<.05$). It contrasts with the findings of Ashcroft et al,[13] which reported that the weight and skeletal age were less than those of controls at all the ages (12 to 21 years) they studied.

It has been argued that most SCD patients, irrespective of their social class, were light in weight and that growth delays occur in both symptomatic and asymptomatic individuals.[17] Socioeconomic class did not influence these parameters in this study. This is understandable because class differences are closing up in our environment due to economic problems.

Findings on the head circumference in this study contrast with that of a previous study,[23] which found a higher value in sicklers due to skull bossing. The mean value in sicklers (52.1 cm±2.5) is lower than in the controls (52.5 cm±2.4), though it is not statistically significant, which rules out skull bossing in SCD subjects in this study. The reduced mean head circumference in the lower socioeconomic class of the SCD subjects might be associated with poor nutrition and a low level of health care in this group. The lower mean MUA found in this study agrees with the previous study by Oguntoye.[18] It has been attributed to reduced skinfold thickness caused by malnutrition[24] and chronic illness.[12] In this environment, the children suffer mostly from malaria, which further depletes red cell count and aggravates malnutrition. Where the affected children are properly nourished, adequate preventive care is put in place and prompt medical care is given, the deficiencies in these health parameters will be abolished.

Conclusions

1. Maxillary prognathism and Class II malocclusion were more prevalent in SCD patients than in control subjects.
2. There was no significant difference in the height and weight values between the two groups except at the age of 18 years, when there was reduction in weight of the SCD group.
3. The MUA circumference of SCD patients was significantly lower than that of control subjects.

Acknowledgments

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ABSTRACT OF THE SCIENTIFIC LITERATURE

STIMULATED PAROTID SALIVARY FLOW RATE IN PATIENTS WITH DOWN SYNDROME

The purpose of this prospective study was to compare the parotid gland salivary output of persons with Down syndrome to healthy individuals. Saliva was collected using a parotid salivary gland cup from 39 subjects (ranging in age from 11 to 62 years) with Down syndrome and from an age and gender matched control group. The authors found that the Down group demonstrated a significantly lower salivary secretion rate compared to the control group. Also, within the Down group, older individuals had a lower flow rate than younger patients and institutionalized individuals had a lower flow rate than those living at home, although these differences were not significant.

Comments: The decreased salivary flow rate increases the importance of establishing a sound prevention plan in these individuals. **MM**

Address correspondence to Dr. Stella Chaushu at drshaush@netvision.net.il

Chaushu S, Becker A, Chaushu G, Shapira J. Stimulated parotid salivary flow rate in patients with Down syndrome. *Spec Care Dentist*. 2002; 22:41-44.

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