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A COMPARATIVE STUDY ON THE SUBNASALE TO GNATHION DISTANCE BETWEEN SICKLE CELL ANEMIA CHILDREN AND NORMAL GROWING CHILDREN IN THE PORT HARCOURT OF NIGERIA

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ABSTRACT

Sickle cell anemia is an inherited blood disorder that arises from gene mutation. This study is designed to determine the subnasale to gnathion distance in children with sickle cell anemia and non sickle cell children in Port Harcourt with age range between 3-18 years. This study was carried out at the University of Port Harcourt Teaching Hospital UPTH (Sickle Cell Clinic). The subnasale to gnathion distance of this sickle cell children and normal growing children were measured from the columella and the upper lip to the lowest point in the lower part of the mandible in the median plane with a sliding caliper. At the end of the study data was collected and tabulated. The data showed that there was a significant difference (P<0.05) in the subnasale to gnathion distance for both sickle cell anemia children and normal growing children of both sexes.

Key words: Sickle cell, non sickle cell, Subnasale- gnathion, Nigeria.

INTRODUCTION

Sickle cell anemia is also known as meniscocytosis or sicklemia, it is an inherited blood disorder that arises from gene mutation. Sickle cell disease is caused by the inheritance of homozygous haemoglobin S (HbSS) or the compound heterozygosity for HbS and HbC (HbSC) or a beta thalassemia, as a result affected hemoglobin molecules have a tendency to stick to one another forming abnormal strands of hemoglobin within the red blood cells. The cell that contains these strands becomes stiff and elongated-sickle-shaped [1].

Only homozygous (HbSS) inheritance of the gene is associated with most severe manifestation while heterozygous (HbAS) individuals or carriers of the trait show no apparent ill effects except under stressful conditions, but show an increased resistance to certain types of malaria. This selective advantage results in a higher occurrence of sickle cell anemia in regions of malarial endemicity such as tropical Africa, India and the Mediterranean [2]. A child who has inherited the sickle cell gene from only one parent will not develop the disease, but will have sickle cell trait. Sickle cell anemia primarily affects people with African, Mediterranean, Middle Eastern and Indian ancestry [3]. The sickling of red blood cells in the absence of oxygen is caused by a change in the hemoglobin molecule structure.

Sickle cell trait is said to occur in 8% of black population in the United States [4]. It is estimated that sickle cell anaemia is directly or indirectly responsible for 8 - 10% of early childhood mortality in much of tropical Africa. Life expectancy is shortened, with studies reporting an average life expectancy of 42 and 48 years for males and females, respectively [5].

The study of normal, abnormal growth and development of children has become an important part of practice [6]. It has been reported that sickle cell anaemia produces stunted growth which usually begins in the second half of the first decade of life and that affected children present with sub-normal weight and height for age measured values [7]. Morphometric parameters such as head, chest and mid-thigh circumference are also useful in evaluating the effect of chronic multi-system diseases such as sickle cell anaemia on the growth pattern of children [8].

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The subnasale, a frequently used reference point is located at the junction of the collumella and the upper lip, while gnathion which is an anatomical landmark at the most inferior border of the mandible in the midsagittal area is located where a line tangent to the pogonion intersects a line tangent to the menton.

The purpose of this study is to determine if sickle cell disease has an effect on the subnasale to gnathion distance and to provide a baseline data of subnasale to gnathion distance between sickle cell and non sickle cell children in Port-Harcourt.

MATERIALS AND METHOD

A total number of 50 patients of both males and females, who are between the ages of 3-18 years that are homozygous for sickle cell at the University of Port Harcourt Teaching Hospital, Port Harcourt, Nigeria, and 250 non sickle cell children from University Demonstration Primary School and Lobo Comprehensive College all in Port Harcourt were recruited for this study. Their subnasale to gnathion distance were measured and recorded on the data sheet.

Measurements

(a.) Measurement of the subnasale-to-gnathion distance was carried out with a sliding caliper by using the method previously described by *Farkas and Lindsay* [9]. The subnasale (sn) is a frequently used reference point that is located at the junction of the columella and the upper lip while the gnathion (gn) is the most inferior midline point

on the mandible, located where a line tangent to the pogonion intersects a line tangent to the mention.

(b)The nasal measurement was done with a sliding caliper by using the method previously described by Farkas and Lindsay [9]. To determine the nasal length, measurement was taken of the distance from a point approximately onehalf centimeters above the area that is in the centre between the eyes or the point of intersection between the frontonasal suture and the midsagittal plane (nasion) to the point where the nasal septum merges with the skin of the upper lip (nasospinale). This is also called the nasion (n) nasospinale (ns) height. The nasospinale is also the point where a line drawn between the inferior most points of the nasal (piriform) aperture crosses the midsagittal plane. This point is not necessarily located at the tip of the nasal spine. The maximum breadth of the nose was measured at a right angle to the nasal height from ala (al-al) to ala. The nose breadth is the distance between the two most lateral points on the wings of the nostrils. These measurements are taken in the absence of any form of facial expression which can alter the size and position of the nose. The nasal index was calculated using the formula: maximum breadth of the nose/ maximum length of nose x 100

Statistical Analysis:

All data were analyzed statistically using the T-test.

RESULTS

The result of this study is as shown in the tables below.

Table 1: Showing the subnasal to gnathion distance of male sickle cell anemia children and non-sickle cell children between 3-18 years. There was a significant difference in the subnasale to gnathion distance between the sickle cell children and non sickle cell children.

Sickle cell				Non sickle cell		
Age group	(years)	Frequency	Mean±sd	Frequency	Mean±sd	
3-6		14	23.21±187	56	26.58±3.76	
7-10		19	23.98±0.46	67	24.44±1.8	
11-14	1	8	23.17±1.03	63	26.46±1.38	
15-18	3	9	25.69±2.6	63	29.16±1.3	
Mean±SD(mm)	24.01±1.49	26.66±1.74	(P<0.05)			

Table 2: Showin	g the subnasale t	to gnathion	distance of fe	male sickle ce	ell anemia	children a	nd non-	sickle ce	ll children:
between 3-18 yea	rs. A significant	difference w	as observed l	oetween the si	ckle cell aı	nd non sick	kle cell ch	nildren.	

Sic	Non sickle cell			
Age group (years)	Frequency	Mean±sd	Frequency	Mean±sd
3-6	4	16.79±-2.71	29	23.27±2.76
7-10	9	23.08 ± -2.78	27	25.13±2.54
11-14	3	11.7±1.52	28	26.40±2.39
15-18	4	23.75±3.3	24	27.15±2.29
Mean±SD (mm) 18.83±2	.57 25.48±2.50	(P<0.05)		

DISCUSSION

In this study Sickle cell disease has been viewed as a disease that is caused by the inheritance of homozygous haemoglobin S (HbSS) or the compound hetero

zygosity for HbS and HbC (HbSC) or a beta thalassemia which was in line with work done by one the researchers [2] *reporting that* homozygous (HbSS) inheritance of the

gene is associated with most severe manifestation while heterozygous (HbAS) individuals or carriers of the trait show no apparent ill effects except under stressful conditions. Morphometric parameters such as head, chest and mid-thigh circumference are also useful in evaluating the effect of chronic multi-system diseases such as sickle cell anaemia on the growth pattern of children [8].

The higher value observed in the subnasale to gnathion distance of the sickle cell disease groups shows that prognathic maxillary profile is more prevalent in sickle cell anaemia (SCA) subjects than the controls. A significant difference was observed in the subnasale to gnathion distance of both the male and female sickle cell children and male and female non sickle cell children [10]. It has been earlier documented that, like other haemolytic anaemia, children with sickle cell anaemia develop a characteristic facial appearance which can be used to describe them in this environment. These characteristics include frontal bossing, prominent maxilla which exposes their teeth, depression of the bridge of the nose and malocclusion of the teeth. This profile is expected to be more prominent in the age group of 3-18 years as evidenced in this study as it revealed a statistical significant difference in comparison to the normal growing children. This finding does not correlate with that of Farkas and James [11].

CONCLUSION

The study therefore, determines some of the craniofacial changes in sickle cell anaemia in comparison to the healthy children in Port-Harcourt. This study could be subjected to further investigation due to its relevance in prediction/clinical anthropometry, maxillofacial surgery and Forensic Science.

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