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Study of physical growth affected by sickle cell diseases

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Abstract
Anthropometric measurements were used to study the physical growth of 294 sickle cell disease (SS) children and compared with 180 normal (AA) children from the Nagpur district of Maharashtra. Both sickle cell disease male and female children were shown to have statistically significant lower weights, heights, sitting heights, hand length, foot length, interacromial diameter and intercristal diameter as compared to normal children with comparable sex and ages. This study concluded that the growth of sickle cell disease children was definitely affected by the disease process. Children with sickle cell disease were shorter, weigh less, and had less transverse diameters.

Keywords: Physical growth, sickle cell disease, hand length, foot length, interacromial diameter and intercristal diameter

Introduction
Anthropometry is concerned with the measurement of the variation of the physical dimensions and the gross composition of the human body at different age levels. It is one of the important parameters for assessment of growth and development and nutritional status of the children. Sickle cell has great effect on the stature, various anthropometric measurement, age, growth and development of the human body.

In 1910, Herrick first reported a case with "peculiar elongated and sickle cell shaped red blood corpuscles" in a case of severe sickle cell anaemia, so called as 'Herrick's anaemia'. Sickle cell anaemia is a serious disease in which the body makes sickle shaped red blood cells. "Sickle-shaped" means that the red blood cells are shaped like a "C".

Normal red cells are disc-shaped and look like doughnuts without holes in the centre. They move easily through blood vessels. Red blood cells contain the protein haemoglobin. This iron-rich protein gives blood its red colour and carries oxygen from the lungs to the rest of the body. Sickle cell contains abnormal haemoglobin that causes the cells to have a sickle shape.

Sickleshaped cells don't move easily through blood vessels. They are stiff and sticky and tend to form clumps and get stuck in the blood vessels. The clumps of sickle cells block blood flow in the blood vessel that leads to limbs and organs. Blocked blood vessels can cause pain, serious infection and organ damage.

Sickle cell anaemia affects almost all systems of the human body. It retards the growth and development, provides unequivocal evidence of impairment in various anthropometric measurements e.g. Height, Weight, Body fat, skeletal maturation, delayed puberty [1-3] Thus sickle cell anaemia acts as a great retarder of human body. This genetic disease is autosomal recessive and hence seen in homozygous individuals. In heterozygous cases, it produces only "sickling" and not the disease. Normal person have haemoglobin A; while in this disease haemoglobin S is found.

In haemoglobin A, glutamic acid (Cytosine-thymine-Cytosine) is seen in Beta chain of haemoglobin at sixth place while in haemoglobin S, Valine (Cytosine-adenine-Cytosine) replace glutamic acid at sixth place [1]. Genetic disorders like Mongolism, Turner's syndrome and Klinefelter's syndrome produce changes in physiognomy, typical to each one of them. The importance of this condition is two-fold, firstly, that this disease is more commonly encountered in Mahar community and secondly that the erythrocytes in this disease are less parasitized by plasmodium, offering resistance to Malaria.[1]

Study of physical growth of sickle cell disease patients had been carried out but data for skeletal age of sickle cell disease patients is not so informative in Indian subjects.

Taking into consideration the above scenario the present project aims to find influence of sickle cell disease on growth of patients by studying anthropometry.

Materials and Method
The present study consists of 294 sickle cell disease (SS) and 180 normal (AA) children in the age group of 5-20 years were examined. This included, 136 male (mean age years) and 158 females (mean age years) with SS and 73 males (mean age years) and 107 females (mean age years) with AA.

The sample was collected from patients who attended the sickle cell OPD or were admitted in the medicine, paediatrics ward of our college, who were willing to participate in the study and sign the informed consent form. The subjects with any obvious congenital or acquired deformity of spine, extremity were excluded from the study. The diagnosis was confirmed by peripheral smear, sickling test and paper electrophoresis.

The study was prospective, non-invasive observational type of study, carried out from January 2015 to December 2015, after obtaining permission from the Institutional Ethical Committee of our institute and from MUHS, Nashik University with the following inclusion and exclusion criteria.

Inclusion Criteria
1. Students of nearby school between 5-20 years of either sex.
2. Patients attending Sickle Cell OPD between 5-20 years of either sex
3. Students and patients willing and signing the written consent form participate in the study. In case of minor

consent was obtained from parent/ guardian of the patients and the students.

Exclusion criteria

1. Subjects below 5 years and above 20 years
2. Subjects having a major medical illness
3. Patients those having skeletal deformity
4. Any obvious congenital or acquired deformity of spine, extremity
5. Subjects having endocrine problems like thyrotoxicosis, dwarfism, rickets, gigantism, Type I DM.
6. Pregnant female
7. Patient of sickle cell disease with other genetic or chromosomal abnormalities
8. Patient of sickle cell disease with congenital heart disease or any congenital anomalies

Consent of patients

We obtained the written consents of the patients attending the SCD OPD by free will through explaining to them before proceeding. The finding of the subjects and controls were noted and entered in the performa. Subjects were measured wearing light clothing and shoes were removed. Anthropometric measurements were taken following the standard techniques. Height was measured using an anthropometer with the subject standing erect with heels together.

Weight measurement was taken on weighing machine, measuring in kilograms up to an accuracy of 100 grams. The zero error was corrected before each measurement. Sitting height was measured with an anthropometer while subject sitting on a horizontal surface, with head in eye-ear plane and his/her trunk stretched to the maximum. Hand length, foot length, interacromial diameter, intercristal diameter measured with the help of spreading calliper. All the above measurements were taken three times and their mean value was taken as a final measurement.

Statistical Analysis

All parameters in sickle cell disease patients were compared with parameters of control by unpaired t-test. The difference was considered significant if the p value was less than 0.05. Data was analyzed on statistical software Graph Pad Prism 5.01.

Results: **- Table No.-1**

Anthropometric measurement	Group I (AA) (n=180)		Group II (SS) (n=294)		
	Male(n=73)	Female(n=107)	Male(n=136)	Female(n=158)	
	Mean± SD	Mean± SD	Mean± SD	Mean± SD	P value
Height (cm)	136.3±19.89	141.2±18.50	125.2±15.72	128.3±16.48	P < 0.0001 Significant (P < 0.05)
Weight (kg)	32.18±12.10	34.27±11.53	22.58±7.745	24.28±8.840	
Sitting Ht (cm)	69.73±8.930	71.38±8.849	65.47±7.009	66.38±8.106	
Hand length (cm)	15.55±2.505	15.74±2.249	13.89±1.820	14.04±1.791	
Foot length (cm)	20.57±2.638	20.80±2.435	18.81±2.299	18.94±2.635	
IA (cm)	27.64±4.125	28.45±3.887	25.00±3.290	25.16±3.224	
IC (cm)	20.14±4.055	20.93±3.701	17.89±2.965	18.60±3.598	

Table 1 shows the results of anthropometric measurement of SS and AA children. Sickle cell disease children (both male and female) showed statistically significant lower values of all the measurement as compared to normal children of the same age and sex group (Fig.1). There was no significant difference between SS males and females for all measurement but female showed less values. Age wise distribution of different anthropometric measurement showed a significant decreased mean value of all measurement in sickle cell disease children as compared to the normal children.

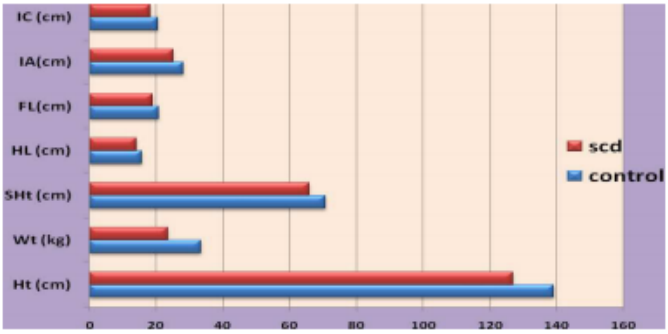

Fig 1: Graphic representation of comparative averages of anthropometric parameters

Discussion

Sickle cell disease, a condition present in Indian population and usually considered to be a clinically benign. However, there is evidence to indicate that the pathophysiology is variable, ranging from a benign to relatively severe clinical manifestations [5]. Although it is generally believed that sickle cell disease has an adverse effect upon the physical growth and development, and however, published data on this aspect from India is meager. Present study deals with the growth and development of sickle cell disease subjects.

294 sickle cell disease and 180 controls were selected and observation regarding their height, weight, sitting height, length of hand, length of foot, inter-acromial diameter, and inter-cristal diameter were recorded. It has been shown that as a group, children with sickle cell disease weigh less and are shorter than the comparable normal controls. Several studies, Winsor et al 1944 [6], Jimenez et al 1966 [7], McCormak et al 1976 [3], Kate 1977 [1], Michael et al 1983 [8], Platt et al 1984 [9], Aschroft et al 1972 [2], Barden EM [10]. (2002), Cepeda ML [11]. (2000), Ebomoyi et al [12]. (1989), Erin L. Bennett [13]. (2011), Hiernaux [14]. (1979), Melissa Rhodes [15]. (2009), Mitchell MJ [16]. (2009), Mukherjee et al [17]. (2004), Saqladi et al [18 (2010), Zemel BS [19]. (2007) have shown that children and adolescent with sickle cell disease have impaired growth as compared to normal controls.

But Aschroft et al 1972 [2] found that the increase of stature during adolescent. Sitting height of sickle cell disease patient measured less as compared to normal controls but McCormack et al (1976) [3]. Found lower values on measures of growth except sitting height. Oredugba et al (2002) [20] found that at the age of 18 years, the weight of sickle cell disease group was significantly less than that of the control group. Patey et al (2002) [21] concluded that the SCD children were taller but had similar weight and BMI to the controls.

But Silva et al (2002) [22] found that the anthropometric measurements did not deteriorate significantly in children with HbSC., Hand length and foot length of sickle cell disease patient also shown less values as observed by Kate (1977) [1].

Similarly our studies reported significant decrease in interacromial and intercristal diameter as found in study of McCormack et al (1976) [3], Kate (1977) [1] Growth delay starts in early childhood but becomes more apparent during

adolescent when the growth spurt of normal children separates them from the patients with sickle cell diseases.

A striking consistent pattern of growth observed in sickle cell disease, shows patients are shorter, weight less and have delay in skeletal age. It is believed that anaemia plays a role in the pathophysiology of sickle cell disease. With respect to physical growth, it has not been determined how anaemia affects either specific organ function or over-all cellular metabolism sufficiently to result in growth retardation.

By multiple transfusions of normal blood anaemia can be corrected and the number of cells capable of sickling can be reduced to clinically insignificant levels. Wolman[23] noted that β-thalassemia patients treated with intensive transfusion therapy appeared in better health and their growth was closer to normal than those transfused only when the hemoglobin level had fallen to low levels. Similar observation were made by Kattamis et al [24] and they concluded that transfusion constitute the treatment of choice for patient with homozygous β-thalasemia, if normal growth is ensured.

Anaemia was found to be very common in our sickle cell disease patients and their hemoglobin levels varied from 3.5 g/dl – 10 g/dl. Although the growth of these sickle cell disease children could be maintain at normal levels through repeated transfusion, however this would not be a feasible therapeutic measure, because they could develop transfusion reactions. Nevertheless, these children with sickle cell disease have a decreased capacity for supplying oxygen to tissues; hence, they may be benefited by having less tissue for oxygenation [5]

Conclusion

Sickle cell disease patient have acute or chronic vaso-occlusion which may lead to poor nutritional status. Even without vasoocclusion patients with hemolysis have lower hematocreit, more marrow, cardiovascular compensation and large caloric requirement. Combinations of these factors affect growth which is consistent with constitutional delay. It can be concluded from the present study that the growth of sickle cell disease in children is significantly affected by the disease process.

Children with sickle cell disease are shorter. They also have less weight and less transverse diameters. It seems likely that several factors may be implicated in different aspects of growth abnormalities seen in children and adults with SS disease. Hence this study indicates a further need for full scale investigation of longitudinal aspect of growth and quantitative assessment of protein and caloric intake of children with clinical and hematological indices of sickle cell disease.

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