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Study of the sickle cell disease affecting physical growth in the children

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Abstract
Sickle cell anaemia is a multisystem disease associated with end-organ damage, which can adversely affect the growth anddevelopment of children. There is no enough data on the effect of this disease on the growth and development of children.
The study was conducted in 25 Sickle cell diseases & 25 normal childrens.
Both sickle cell disease male and female children were shown tohavestatistically significant lower weights, heights, sitting heights, interacromial diameter and intercristal diameter ascompared to normal children.
The further investigations are needed in this study for growth and its quantitative relation with the proteins, caloric intake and haematological index of the children.Sickle cell anaemia is not causing significant adverse effect on the growth of children; but it has a transient negative effect onthe growth of primary school going children.

Keywords: sickle cell disease, physical growth, physical growth etc.

Introduction
Sickle-cell disease (SCD) is a group of blood disorders typically inherited from a person's parents. The most common type is known as sickle-cell anaemia (SCA). It results in an abnormality in the oxygen-carrying protein haemoglobin (haemoglobin S) found in red blood cells. This leads to a rigid, sickle-like shape under certain circumstances. Problems in sickle cell disease typically begin around 5 to 6 months of age. A number of health problems may develop, such as attacks of pain ("sickle-cell crisis"), anemia, swelling in the hands and feet, bacterial infections, and stroke. Long term pain may develop as people get older. The average life expectancy in the developed world is 40 to 60 years.
Sickle-cell disease occurs when a person inherits two abnormal copies of the haemoglobin gene, one from each parent. This gene occurs in chromosome 11. Several subtypes exist, depending on the exact mutation in each haemoglobin gene. An attack can be set off by temperature changes, stress, dehydration, and high altitude. A person with a single abnormal copy does not usually have symptoms and is said to have sickle-cell trait. Such people are also referred to as carriers. Diagnosis is by a blood test and some countries test all babies at birth for the disease. Diagnosis is also possible during pregnancy.
Sickle cell anemia (SCA) is a genetic blood disorder caused by abnormal inherited hemoglobin. The abnormal hemoglobin causes distorted or sickle-shaped red blood cells i.e. the red blood cells are shaped like a crescent. It is the most common form of sickle cell disease (SCD). It is also known as Hemoglobin SS disease, Hemoglobin S disease, HbS disease or sickling disorder due to hemoglobin S.
The highest frequency of Sickle Cell Disease is found in tropical regions, particularly Sub-Saharan Africa, India and the Middle-East. Migration of substantial populations from these high prevalence areas to low prevalence countries in Europe has dramatically increased in recent decades. The prevalence of Sickle Cell Anemia is highly common in the tribal belt of Central and Southern India. The public health implications of Sickle Cell Anemia are significant leading to poor quality of life, lower life expectancy and higher rates of infant mortality. Sickle cell disease (SCD) with an estimated 5,200 live births each year is a major public health problem in India. Although SCD has been described in India in numerous ethnic groups, it is most prevalent. Prevalence of Sickle Cell gene is 5 to 34 % in scheduled tribes, who have a high prevalence of socio-economic disadvantage and are frequently medically underserved.
India has also a very huge populations of tribal community about 18 crore and expected to have 1.80 crore sickle cell trait and 14 lakhs of sickle cell disease. These show the big burden on the public health of India.
SCD is a serious group of conditions which are inherited (genetic). It affects the red blood cells in the blood. Sickle cell anaemia is the name of a specific form of SCD in which there are two sickle cell genes. Sickle cell disease (SCD) is one of the most commonmonogenic disorders globally with an autosomal recessive inheritance [1]. James Herrick, a physician first described the characteristic sickle shaped red cells in a medical student from Grenada in 1910. Linus Pauling and his colleagues showed that sickle haemoglobin (HbS) had an altered electrophoretic mobility and they were the first to define it as a molecular disease in 1949. A few years later in 1957, Vernon Ingram discovered that sickle haemoglobin resulted from a single amino acid substitution in the haemoglobin molecule [2, 3]. The disease results from a single base A>T mutation in the triplet encoding the sixth residue of the β-globin chain, leading to a substitution of valine for glutamic acid and the abnormal haemoglobin S (HbS).
The primary pathophysiology is based on the polymerization of deoxyHbS with formation of long fibers with in the RBCs causing a distorted sickle shape which eventually leads to increased haemolysis and vaso-occlusion of sickle red cells. However, the clinical presentation of SCD patients is extremely variable and there are several events that may trigger

vasoocclusion. Recent work has shown the importance of red cell dehydration, abnormal adhesion of RBCs to the vascular endothelium, imflammatory events, and activation of all the cells in the vessel and abnormalities of nitric oxide metabolism in the pathophysiology of this multi-organ disease [4].

Earlier reports have shown that American Black children with sickle cell disease were shorter with lower weights and generally thinner body build than normal children [5] In India, the βS gene is prevalent especially in the tribal populations and the prevalence rate varies from 0 - 40% in different population groups [6] Several workers have reported the molecular basis of sickle cell disease particularly with reference to its milder clinical manifestations as compared to the Afro-Carribean counterpart [7, 8].

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.

Materials and Methods [9, 10].

The study was planned into the 25 sickle cell disease and 25 normal children's were evaluated. All the childrens are from the Anugrah narayan magadh medical college and hospital. The study includes about the 25 male and female children's. visited the Pediatric Out-Patient Department (OPD) and in-patient department (IPD) of Anugrah narayan magadh medical college and hospital were considered in the study. All the patients are informed consents.

Group A: 25 sickle cell disease children.
Group B: 50 normal children's

Inclusion Criteria

1. Children's of either sex.
2. Age between 5-15 years
3. Patients referred in IPD & OPD both departments.

Exclusion Criteria

1. Children's below 5 years & new-born.
2. Patient of sickle cell disease with other genetic or chromosomal abnormalities.
3. Patient having other medical diseases.
4. Children's having endocrine abnormalities.

The anthropometric measurements were taken following the standard techniques. Weight measurement was taken on a balance scale. Height was measured using an anthropometer with the subject standing erect with heels together. Skin fold thickness was measured using a Harpenden skinfold caliper. Other measurements were taken included sitting height and mid arm circumferences. Upper/ lower segment (U/L) ratio and body mass index were expressed as sitting height /distance from the top of public symphysis to the floor and weight in kg / height in metre respectively.

Results and Discussion

Table 1 shows the results of the anthropometric measurements of age and sex matched Sickle cell and Normal children.

Table 1

Anthropometric measurement	Group A: 25 sickle cell disease children	Group B: 25 normal children's
Height (cm)	108.6 ±12.4	123.3±13.5
Weight (kg)	14.2±4.2	19.2±3.22
Sitting Height (cm)	55.5±6.11	64.4±8.2
Interacromial diameter (cm)	25.50±3.60	27.6±2.7
Intercristal Diameter (cm)	18.5±2.8	22.3±2.3

Sickle cell disease children showed statistically significant lower values of all the measurements.

Sickle cell disease, a condition present in Indian populations and usually considered to be a clinically benign. However, there is evidence to indicate that the pathophysiology is variable, ranging from a benign to are latively severe clinical manifestations. Although it is generally believed that sickle cell disease has an adverse effect upon the physical growth and development, however, published data on this aspect from India is meagre.

In the present study, 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 from the United States, Jamaica, Italy and Nigeria have shown that children and adolescents with sickle cell disease have impaired growth as compared to normal controls.

Growth delay starts in early childhood but becomes more apparent during adolescence when the growth spurt of normal children separates them form the patients with sickle cell disease.

Delayed sketetal maturation and adolescent growth spurt have also been reported [11]. The growth deficittendsto be greater in weight than in height and is more severe in patients with sickle cell anemia and S-β0thalassemiathan in those with HbSC disease and Sβ+ thalassemia [12].

It is believed that anemia plays a role in the pathophysiology of sickle cell disease. With respect to physical growth, it has not been determined how anemia affects either specific organ function or over-all cellular metabolism sufficiently to result in growth retardation.

Conclusion

Sickle cell diseases patients are having vasoconclusion leads to the poor nutritional status. Also due to vasoconclusion there arises different condition like lower hematocreit and more caloric need. So due to these type of factors may affect the growth and development. These are relevant to the findings in both group of childrens. The present study denotes that the patients with sickle cell diseases are shorter in height. Also have less weight and Interacromial & Intercristal diameter. The further investigations are needed in this study for growth and its quantitative relation with the proteins, caloric intake and haematological index of the children.

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