Physical growth of children with sickle cell disease

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Introduction

Sickle cell disease is one of the commonest single gene disorders in man and has a widespread distribution in different parts of the world with variable clinical manifestations. An important clinical issue requiring further clarification is the effect of this abnormal hemoglobin on the physical growth and development of children with sickle cell disease. 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. In India, the βS gene is prevalent especially in the tribal populations and the prevalence rate varies from 0 - 40% in different population groups. 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. However, there is paucity of data on physical growth and development of sickle cell disease children in India.

Our population survey in Nagpur district of Maharashtra revealed a high prevalence of βS gene (23%) among the Mahar community. Fifty eight children with sickle cell disease were identified during this population screening. The clinical manifestations of these cases were severe and this data has been published elsewhere. In this report, we present the study on physical growth status of sickle cell disease children by using some of the anthropometric measurements.

Materials and Methods

All children in this study belonged to the Mahar group from Nagpur district of Maharashtra. A total of 58 sickle cell disease (SS) and 86 normal (AA) children in the age group of 2 - 14 years were examined. This included, 38 boys (mean age 7.8 years) and 20 girls (mean age 8.7 years) with SS and 45 boys (mean age 8.5 years) and 41 girls (mean age 9.7 years) with AA. The diagnosis
of sickle cell disease was made on the basis of cellulose acetate electrophoresis at pH 8.9 as described earlier.7

Subjects were measured wearing light clothing and shoes were removed. The anthropometric measurements were taken following the standard techniques.8 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^2 respectively. Statistical analysis was done by using students t-test.

Results

Table 1 shows the results of the anthropometric measurements of age and sex matched SS and AA children. Sickle cell disease children (both male and female) showed statistically significant lower values of all the measurements except U/L segment ratio as compared to normal children of the same age and sex groups. There was no significant difference between SS males and females for all the measurements. BMI values were also found to be significantly lower among the SS male and female children than the normal children. Age wise distribution of different anthropometric measurements showed a significant decreased mean values of all the measurements except U/L ratio in the age group of 11-14 years than the earlier age group in sickle cell disease children as compared to the normal children.

Discussion

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 a relatively severe clinical manifestations.6 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 meager.

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.9 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 skeletal maturation and adolescent growth spurt have also been reported.10 The growth deficit tends to be greater in weight than in height and is more severe in patients with sickle cell anemia and S-β^0 thalassemia than in those with HbSC disease and Sβ^+ thalassemia.11

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. By multiple transfusions of normal blood anemia can be corrected and the number of cells capable of sickling can be reduced to clinically insignificant levels. Wolman12

Table 1: Anthropometric measurements of sickle cell disease and normal children

<table>
<thead>
<tr>
<th>Anthropometric Measurements</th>
<th>Males</th>
<th></th>
<th></th>
<th>Females</th>
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<th></th>
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</thead>
<tbody>
<tr>
<td></td>
<td>SS (n = 38) Mean ± SD</td>
<td>AA (n = 45) Mean ± SD</td>
<td>t values</td>
<td>SS (n = 20) Mean ± SD</td>
<td>AA (n = 41) Mean ± SD</td>
<td>t values</td>
</tr>
<tr>
<td>Weight (kg)</td>
<td>17.8 ± 7.3</td>
<td>22.0 ± 9.2</td>
<td>8.14*</td>
<td>17.7 ± 8.2</td>
<td>22.8 ± 10.1</td>
<td>6.27*</td>
</tr>
<tr>
<td>Height (cm)</td>
<td>115.0 ± 20.5</td>
<td>124.4 ± 22.5</td>
<td>10.00*</td>
<td>114.9 ± 19.1</td>
<td>123.6 ± 21.2</td>
<td>7.27*</td>
</tr>
<tr>
<td>Sitting height (cm)</td>
<td>59.2 ± 9.8</td>
<td>65.2 ± 9.9</td>
<td>9.43*</td>
<td>58.7 ± 10.5</td>
<td>65.0 ± 10.2</td>
<td>7.46*</td>
</tr>
<tr>
<td>U/L Ratio (cm)</td>
<td>1.0 ± 0.2</td>
<td>1.1 ± 0.1</td>
<td>0.62</td>
<td>1.0 ± 0.2</td>
<td>1.1 ± 0.2</td>
<td>0.66</td>
</tr>
<tr>
<td>Mead arm circumferences (cm)</td>
<td>13.8 ± 1.3</td>
<td>14.8 ± 2.1</td>
<td>3.62*</td>
<td>14.3 ± 2.5</td>
<td>15.3 ± 2.8</td>
<td>2.22**</td>
</tr>
<tr>
<td>Skin fold thickness (mm)</td>
<td>6.8 ± 1.9</td>
<td>8.0 ± 2.4</td>
<td>3.63*</td>
<td>6.2 ± 2.4</td>
<td>7.8 ± 3.3</td>
<td>3.53*</td>
</tr>
<tr>
<td>Body mass index</td>
<td>12.4 ± 1.3</td>
<td>13.6 ± 1.6</td>
<td>4.20*</td>
<td>12.6 ± 2.0</td>
<td>14.1 ± 2.2</td>
<td>3.80*</td>
</tr>
</tbody>
</table>

*P<0.001; **P<0.05 n = Number of individual studied
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 Hb had fallen to low levels. Similar observations were made by Kattamis et al\textsuperscript{13} and they concluded that intensive transfusions constitute the treatment of choice for patients with homozygous β-thalassemia, if normal growth is to be ensured. Anemia was found to be very common in our sickle cell disease patients and their hemoglobin levels varied from 3.5 g/dl – 10.0 g/dl with a mean of 7.8 ± 1.8 g/dl. Although the growth of these sickle cell disease children could be maintained at normal levels through repeated transfusions, 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.

The body mass index (BMI) is a body mass per unit area and a measure of adiposity of an individual and found to be a good indicator of nutritional status. However, it is difficult to find out whether the inadequacy of nutrients is due to inadequate diet or poor absorption or defective metabolic utilization by an individual. In the present study, it was found that the children with sickle cell disease are under nourished which has been reflected by a significantly lower mean BMI values, mid arm circumferences and skin fold thickness than the normal controls. Similar observations have been reported on adult sickle cell patients from Orissa.\textsuperscript{14} The low BMI in our sickle cell disease children could be due to inadequate food intake because of poor appetite especially during the vasoocclusive crisis.

Hence, this study indicates a further need for full scale investigation of longitudinal aspects of growth and quantitative assessment of protein and caloric intake of the children with sickle cell disease and other hemoglobinopathies.

References