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IMPACT OF DIFFERENT FORMS OF SICKLE CELL DISEASE ON THE GROWTH AND NUTRITIONAL STATUS OF CHILDREN

Paediatrics

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ABSTRACT

Introduction: Sickle Cell Disease(SCD) is a progressively debilitating genetic disease which impacts millions of children worldwide. Most common type of SCD are HbSS, HbAS/sickle cell trait and sickle cell with beta-thalassemia. India accounts for 14.5% of total newborns with SCD worldwide. Population with SCD suffered from chronic form of malnourishment hence resulted in lower weight, height or BMI. **Objective:** The objective of the study was to observe the impact of different forms of sickle cell disease like sickle cell anemia, sickle cell trait and sickle cell anemia with thalassemia on growth and nutritional status of the children with reference to WHO growth standards and to find out that the impact caused by different form of disease is significantly different from each other or not. **Material and Method:** An observational and cross-sectional study was conducted in Index Medical College Hospital and Research Centre, Indore from September 2022 to February 2024. The study enrolled 100 paediatric patients with Sickle Cell Disease between 5 - 18 years age group. The anthropometric variables for all children were recorded and projected on WHO charts 2007 which was expressed in Z-scores and was gender specific. **Results:** About 71% population were classified as HbSS and the rest 15% as HbAS and 14% as HbS–beta-thalassemia genotype. No significant difference was found between the magnitude of growth affected by 3 different forms of sickle cell disease. **Conclusion:** All forms of the sickle cell disease, sickle cell anemia, sickle cell trait and sickle cell with thalassemia suffer from similar proportion of stunting and undernutrition which was severe in its magnitude. There is requirement to launch a study of broader magnitude to screen, diagnose all the form of disease. Difference in the severity of growth retardation was not significant in different form of SCD.

KEYWORDS

Sickle Cell Disease, Nutritional Status, Growth, Children, Z-score

INTRODUCTION

Sickle Cell Disease (SCD) is a progressively debilitating genetic disease which impacts millions of children worldwide. It results from single nucleotide substitution, converting a glutamic acid codon to a valine codon at 6th position of the beta globin chain. This results in changes in physical properties of globin chain leading to hemoglobin S formation which polymerizes during physiological stress causing erythrocyte to take sickle shape. This event leads to vaso-occlusion and hemolytic anemia which play a central role in the clinical complications associated with SCD.[1]

Most common type of SCD are HbSS, HbAS/sickle cell trait and beta thalassemia. When an individual has a pair of sickle gene inherited from both of the parent then it forms homozygous sickle cell disease(HbSS). When an individual possess only one HbS gene then it forms heterozygous condition known as sickle cell trait(SCT) (HbAS).[2] Sickle cell anemia- beta thalassemia is an inherited form of SCD that affects red blood cells both in production of abnormal hemoglobin as well as the decreased synthesis of beta globin chains.

India accounts for 14.5% of total newborns with SCD worldwide.[3] In India, SCD is prevalent among socioeconomically disadvantaged ethnic group, scheduled tribes, scheduled caste and backward castes.[4]

The objective of the study was to observe the impact of different forms of sickle cell disease like sickle cell anemia, sickle cell trait and sickle cell anemia with thalessemia on growth and nutritional status of the children with reference to WHO growth standards and to find out that the impact caused by different form of disease is significantly different from each other or not.

MATERIAL AND METHOD

An observational and cross-sectional study was conducted in the Paediatrics department of a tertiary care hospital during the period of 18 months from September 2022 to February 2024. The enrolled paediatric population was consisted of male and female children between 5 to 18 years of age group with confirmed clinical and laboratory diagnosis of SCD attending OPD, IPD and PICU of the hospital.

Inclusion Criteria

1. Known cases or recently diagnosed cases of sickle cell disease in children of age group 5-18 years of either sex attending OPD, IPD and PICU.
2. Children whose parents gave informed consent to participate by signing in consent form.

Exclusion Criteria

1. Cases of nutritional deficiency anaemia, thalassemia and other haemoglobin abnormalities.
2. Patients with chronic kidney and liver disease.
3. Patients with co-morbidities like congenital or rheumatic heart disease and chromosomal abnormalities.

For growth assessment height, weight and BMI were measured. The anthropometric variables for all children were projected on WHO charts 2007 which was expressed in Z-scores and was gender specific.

HAZ (Height for age Z-score):

This reflected the growth achieved in height while standing at a particular age. This score was used to determine stunting. For a Z-score of <-3SD, stunting was noted as severe. Z-score between -3 to -2SD was noted as mild stunting; -2 to +2 was considered normal and more than +2SD was noted as very tall.

WAZ (Weight for age Z-score):

This reflected body weight in a particular age. This score showed if a child was underweight or severely underweight. Between -3 to -2SD, it was underweight. When this Z –score was < -3SD, it was called severe underweight. For children more than 10 years, WAZ score could not be computed because WHO charts were only for children younger than 10 years of age.

BMIZ (BMI for age):

The BMI for age was calculated from weight and height of child and plotting the given value on WHO chart. Severe underweight was considered for < -3SD; underweight was noted between -3 to -2SD and normal between -2 to +2SD.

The statistical analyses were performed using the Statistical Package SPSS-20. P-value for frequency percentage comparison was calculated. P-value<0.05 was considered statistically significant. The Ethical approval was obtained from the Institutional Ethics Committee.

RESULTS

Out of the study population it was observed that 71% of the study subjects suffered from Sickle Cell Anemia (HbSS)(homozygous variety), 15% suffered from sickle cell anemia with thalassemia and

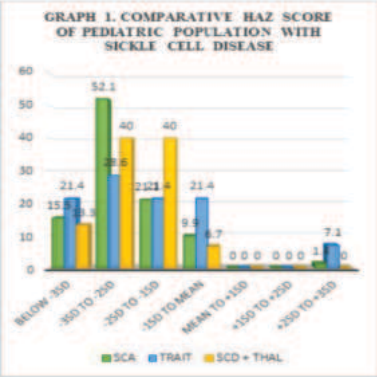
14% suffered from sickle cell trait(HbAS)(heterozygous variety). When the comparison was made in different domains of growth, following results were observed.

HAZ (Height for age Z-score):

In population with SCA and beta thalassemia, nearly 53.5% were stunted out of which 13.3% were severely stunted. In comparison, population with SCA homozygous variety, 67.6% were stunted, out of which 15.5% were severely stunted. In population with sickle cell trait, 50% were stunted, out of which 21% were severely stunted. When compared with WHO standard, significant difference was seen. But when all three of the groups were compared with each other, no significant difference was observed in the heights of the subjects.

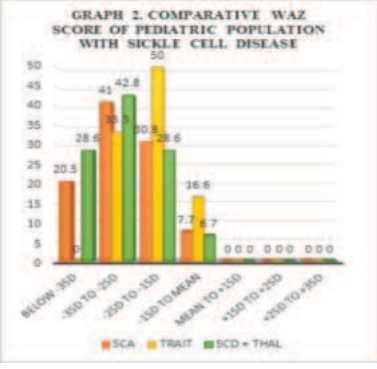
Table 1.nutritional Status of the Pediatric Population with Sickle Cell Disease on the Basis of Mean Z-scores.

S.NO.	VARIABLES (Z-SCORES)	POPULATION (N%) Sickle cell trait	POPULATION(N%) SCA + Thal	POPULATION (N%) SCA	P VALUE
1	HEIGHT FOR AGE (HAZ)				0.233
	❖ BELOW -3SD	21.4	13.3	15.5	
	❖ -3SD TO -2SD	28.6	40.0	52.1	
	❖ -2SD TO -1SD	21.4	40.0	21.1	
	❖ -1SD TO MEAN	21.4	6.7	9.9	
	❖ MEAN TO +1SD	0	0	0	
	❖ +1SD TO +2SD	0	0	0	
	❖ +2SD TO +3SD	7.1	0	1.4	
2	WEIGHT FOR AGE (WAZ)				0.213
	❖ BELOW -3SD	0	28.6	20.5	
	❖ -3SD TO -2SD	33.3	42.8	41.0	
	❖ -2SD TO -1SD	50	28.6	30.8	
	❖ -1SD TO MEAN	16.6	0	7.7	
	❖ MEAN TO +1SD	0	0	0	
	❖ +1SD TO +2SD	0	0	0	
	❖ +2SD TO +3SD	0	0	0	
3	BMI FOR AGE (BMIZ)				0.189
	❖ BELOW -3SD	21.4	26.7	11.3	
	❖ -3SD TO -2SD	14.3	20.0	12.6	
	❖ -2SD TO -1SD	7.1	26.7	28.2	
	❖ -1SD TO MEAN	35.7	26.6	33.8	
	❖ MEAN TO +1SD	21.4	0	11.3	
	❖ +1SD TO +2SD	0	0	2.8	
	❖ +2SD TO +3SD	0	0	0	



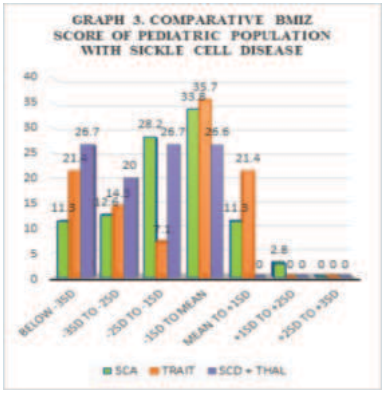
WAZ (Weight for age Z-score):

In population with SCA with beta thalassemia, 71.2% of the population was underweight with 28.6% of them being severely undernourished. This proportion was changed in population with SCA where 61.5% of population was underweight in which 20.5% were severely underweight. This proportion surprisingly changed in population with Sickle cell trait where only 33.3% population was underweighted and no one suffered from severe undernutrition. When compared with WHO standard, significant difference was seen. But when all three of the groups were compared with each other, no significant difference was observed in the weight for age of the subjects in the groups.



BMIZ (BMI for age):

When BMIZ was compared it was observed that in population with SCA with beta thalassemia, 46.7% were underweight, out of which 26.7% were severely underweight. In SCA homozygous population only 23.9% were underweight while 11.3% were severely underweight. However, in population with sickle cell trait 35.7% were underweight, out of which 21.4% were below -3 S.D. When compared with WHO standard, significant difference was seen. But when all three of the groups were compared with each other, no significant difference was observed in the BMIZ scores of the subjects in the groups.



DISCUSSION

Sickle cell disease is a hereditary, autosomal recessive condition which has a chronic, long-lasting effect on the body of the suffering patient. The disease, whether in homozygous or heterogenous form, affects the body in very wide range. The disease affects growth of the body by affecting the growing end of bones in repetitive vaso-occlusive crisis. Patients often end up in hospitals because of infections and other complications which consumes lot of resources. Endocrine and nutritional deficiencies are the other cause which retarded the growth in one or more of the anthropometric indices like BMI, height and weight.[5,6]

Our objective was to measure growth in form of anthropometric measurements like height, weight and BMI of paediatric population with SCD. In our study, WHO growth charts were taken for z-score references. Fadhil et al[7] also defined stunting, wasting and underweight as HAZ, WAZ and BMIZ scores < -2 SD from the median WHO standard.

Kazadi et al[8] noticed that population with SCD suffered from chronic form of malnourishment hence resulted in smaller size, lower weight for height or BMI. Similar findings were seen in our study. Significant proportion of population suffering from all the forms of SCD had growth retardation in some or the other form.

In population with Sickle cell anemia homozygous variety, 15.5% were severely stunted among the 67.6% stunted population. WAZ and BMIZ were also affected as noticed in other studies. It was observed that in WAZ domain, 61.5% of population was underweight in which 20.5% were severely underweight. While BMIZ measurement showed 23.9% of population was underweight out of which 11.3% were underweight.

In population with SCA with beta thalassemia, 71.2% of the population was underweight with 28.6% of them being severely undernourished. 53.5% were stunted out of which 13.3% severely stunted and when BMI of this population was taken,46.7% were underweight and 26.7% were severely underweight.

When population with sickle cell trait was observed, 50% were stunted out of which 21% were severely stunted. BMIZ and WAZ followed similar pattern. When weight for age of sickle cell anemia were observed, 33.3% were underweight but no one suffered from severe undernutrition. In observation of BMIZ of the sickle cell trait population 35.7% were underweight out of which 21.7% were severely underweight.

However when all the three variants of sickle cell disease were compared with each other, it was observed that no significant difference was noted in between the magnitude by which growth of

population of sickle cell anemia, sickle cell trait and sickle cell anemia with thalassemia was affected.

CONCLUSIONS

Prevalence of sickle cell disease in its all forms is high in central India. Nearly all the forms of the sickle cell disease, sickle cell anemia, sickle cell trait and sickle cell with thalassemia suffered from similar proportion of stunting and undernutrition which was severe in its magnitude. Thus there is requirement to launch a study of broader magnitude to screen and diagnose all the form of disease and to find out the means of proper and early interventions to reduce the effect of the disease on the growth component of the disease.

REFERENCES

[1] Menezes AS, Len CA, Hilario MO, Terreri MT, Braga JA. Quality of life in patients with sickle cell disease. Rev Paul Pediatr. 2013;31(1):24-9.
[2] Serjeant GR. The emerging understanding of sickle cell disease. Br J Hematol. 2001;112:3-18.
[3] Colah RB, Mukherjee MB, Martin S, Ghosh K. Sickle cell disease in tribal population in India. Indian J Med Res. 2015; 141: 509-15.
[4] Rao VR. Genetics and epidemiology of sickle cell anemia in India. Ind J Med Sci. 1988; 42(9):218-22.
[5] Cepeda ML, Allen FH, Cepeda NJ, Yang YM. Physical growth, sex maturation, body image and sickle cell disease. J Nat Med Assoc. 2000;92(I):10-14.
[6] Odentundu OI, Chinawa JM, Achigbu KI, Achigbu EO. Body mass index and other anthropometric variables in children with sickle cell anemia. Pak J Med Sci. 2016; 32(2):341-6.
[7] Fadhil RS, Hassan MK, Al-Naama LM. Growth and nutritional status of children and adolescents with sickle cell anemia. Egypt J Haematol. 2020;45:188-96.
[8] Kazadi AL, Ngiyulu RM, Gini- Ehungu JL, Mbuyi- Muamba JM , Aloni MN. Factors associated with growth retardation in children suffering from sickle cell anemia: first report from Central Africa. Anemia. 2017; 2017 Article ID 7916348.