Sinha A., Patra, P.K., Panda P.S. 2024. "Nutritional challenges and determinants in children with sickle cell disease in Chhattisgarh, India." *World Nutrition*, *15*(4), December, 47–51. https://doi.org/10.26596/wn.202415447-51

Research

Nutritional challenges and determinants in children with sickle cell disease in Chhattisgarh, India

Ashish Sinha[1], Pradeep Kumar Patra[2], Prem Sagar Panda[3]

[1]Department of Community Medicine, Pt. JNM Medical College, Raipur, India., [2]Pt Dindayal Upadhyay Health Science and AYUSH University, Raipur, India. [3]Department of Community Medicine, Kalinga Institute of Medical Sciences, Bhubaneswar, India

Keywords: Sickle cell disease, malnutrition, children, dietary supplementation, hospitalization.

https://doi.org/10.26596/wn.202415447-51

World Nutrition 2024;15(4):47-51

Background

Sickle cell disease (SCD) is one of the commonest single gene disorders recognized as a public health priority. Slow growth is common in children with SCD due to the vicious cycle of malnutrition and infection among them. Increasing knowledge that nutritional problems are fundamental to the severity of the disease has produced interest in promoting dietary supplementation for treating these patients.

Objective

The objective of the present study was to assess nutritional status and associated factors in SCD patients registered at the Sickle Cell Institute Chhattisgarh, Raipur, CG, India.

Methods

We reviewed clinical data from documents/secondary records among a cohort of 671 children with SCD 0-18 years oldat the Sickle Cell Institute.

Results

Out of all 671 children included in the study, 380 (56.6%) were male.Underweight, stunting and wasting proportions were 58%, 37.7% and 38.9% respectively. 16% children had chronic malnutrition (low height for age). 62% of the malnourished children were ⩾ 11 years old and38% were ⩽ 10 . 64% of the children who needed hospitalization were also in the older age group. Chronic malnutrition was significantly higher among children with an O Rh+ blood group (39.8%). 26.7% had sought care in hospital before diagnosis either for blood transfusion or for a vaso-occlusive crisis.

Conclusions

A significant proportion of chronic malnutrition was seen among the study population mixed with underweight, stunting and wasting. Older childrenwith an O+Ve blood group had increased risk of hospital admission. Children should get priority for nutritional intervention as adjunct treatment with standard care practice to increase survival chances.

INTRODUCTION

Hemoglobinopathy is a common inherited disorder of red blood cells. Sickle cell disease (SCD) is one of them. The most common gene disorders are sickle cell anemia (HbSS or SCA), hemoglobin SC (HbSC) and hemoglobin Sβ thalassemia (HbSβthal). Patients with SCA suffer most severely (Ministry of Health and Family Welfare, Government of India. 2016*)*. These diseases represent a significant global public health concern, particularly in environments endemic for malaria *(*Ministry of Health and Family Welfare, Government of India. 2023*)*. Morbidity and mortality caused by these pose a public health burden on families and the health sector in a country like India. SCD affects many communities in certain regions, such as Gujrat, Maharashtra, Rajasthan, Madhya Pradesh, Jharkhand, Chhattisgarh, West Bengal, and Odisha innorthern India and in the southern states of Tamil Nadu, Telangana and Kerala *(*Ministry of Health and Family Welfare, Government of India. 2023*)*.

[a] Corresponding author: ashishsinha.md@gmail.com

INTRODUCTION

The carrier frequency of the sickle cell gene varies from 1-35%, suggesting that huge number of people carrying the gene of SCD (Ministry of Health and Family Welfare, Government of India. 2016). SCD is especially prevalent amongst tribal peoples. Hence, an expert committee on tribal health has listed SCD as one of the 10 special problems they suffer from.

The Ministry of Tribal Affairs has chalked out a detailed action plan to ensure that there are no newborns with SCD by 2025 (Ministry of Health and Family Welfare, Government of India. 2023*).* Hydroxyurea (HU) is the only disease modifying drug approved by the FDA for treating SCD (Platt, 2008). However, average life expectancy is still approximately 30 years lower for individuals with SCD than for the general population (Platt, 1994).

Due to the worldwide use of HU, researchers have paid less attention to new studies addressing nutritional deficiencies, which still exist and contribute to slowed growth and development and compromising their quality of life and likely shortening their life span. Dietary recommendations for SCD should include more emphasis on adequate amounts of macronutrients (Hyacinth, 2018). People living with SCD have a relative energy shortage. Nutritional deficiency in SCA is secondary to a marked hypermetabolic state, in turn associated with an increased energy requirement. Dietary supplementation for treating SCD should be considered for these patients (Platt, 2008).

Two types of hemoglobin (Hb) are measured in clinical diagnosis and treatment of SCD. In addition to normal Hb, fetal hemoglobin (HbF, $\propto_2\gamma_2$) is measured. It can inhibit the deoxygenation-induced polymerization of sickle hemoglobin (HbS, $\alpha_2\beta^S{}_2$) that drives the pathophysiology of sickle cell disease (Eaton WA, 1987).

While SCD children living in Chhattisgarh are likely at risk of malnutrition, there has been no evidence available on this issue. Thus, there are no guidelines for their nutritional management. Hence, the present study was conducted to ascertain the extent and determinants of nutritional challenges among the SCD patient registered in Chhattisgarh.

MATERIALS AND METHOD

We reviewed clinical data among a cohort of 671 children 0-18 years with SCD, collecting relevant information of all the patients from documents/secondary data available at Sickle Cell Institute Chhattisgarh, Raipur. The universal sampling method was followed and data collection was done from the patient records. As existing record-based data were collected, permission from an Institutional Review Board was not sought. Anonymization protocols were followed to ensure compliance with ethical standards. Data entry was performed in a Microsoft Excel spreadsheet, followed by analysis conducted with IBM SPSS Statistics for Windows, Version 23 (Released 2015; IBM Corp., Armonk, New York, United States). Data are presented as frequencies and percentages. The chi-square test was done to analyse the association among categorical variables. $P < 0.05$ was taken to be statistically significant.

RESULTS

Out of all 671 children included in the study, 380 (56.6%) were males, 72.1% were from the so-called Other Backwards Class (OBC) while 6.11% were Schedule Tribe (Table 1). Underweight, stunting and wasting were 58%, 37.7% and 38.9% respectively (Table 2). 16% had chronic malnutrition (Table 3). Chronic malnutrition was significantly associated with a Hb concentration of < 10 mg/dl ($P < 0.01$). Chronic malnutrition was significantly higher (39.8%) among the O +Ve blood group in comparison to other blood groups. As shown in Table 4, older children (11-18 Years) were significantly more likely to be chronically malnourished (62.04%) in comparison to younger children (38%) (1-10Years)($P < 0.01$). Among those chronically malnourished, older children (64%) needed hospital care more often than younger children (36%) ($P < 0.01$). The majority (73.9%) were diagnosed with SCD beyond 5 years of age. At time of diagnosis, their most common symptoms were generalized bodyache together with vaso-occlusive crisis (VOC) (28.2%) followed by icterus and pallor (27.7%), and bone and joint pain (2.53%). Only 14.7% were asymptomatic. 26.7% were already in hospital before diagnosis, either for blood transfusion (BT) or for after an episode of VOC. Patients with an Hb level>10mg and fetal hemoglobin (Hb F)>20% were at significantly lower risk of needing hospitalization. Table 5 indicates the pattern of morbidity in the sample children. Table 6 shows the link between children who needed hospitalization and levels of malnutrition. Chronic malnutrition was found to be significantly associated with presence of HbA/HbF (Table 7). Similarly, blood group and age were significantly associated with underweight (Table 8). Other measures of malnutrition were not associated with fetal Hb or blood group.

Table 1. Socio-demographic profile of the children registered at Sickle Cell Institute Chhattisgarh (n=671)

Variables	Categories	Number	%
Sex	Male	380	56.64
	Female	291	43.36
Age	Up to 5	175	26.00
	6-11	291	43.36
	12-18	205	30.64
Type of sickle cell	General	11	01.64
	OBC	493	73.47
	ST	41	06.11
	SC	126	18.78
Age at diagnosis	< 5 Yrs	175	26.00
	> 5 Yrs	496	74.00

Table 2. Nutritional status of children registered at Sickle Cell Institute Chhattisgarh (n=671)

Nutritional Status	Present	%	Absent	%
Underweight	389	57.97	282	42.03
Stunted	253	37.70	418	72.30
Wasted	261	38.90	410	61.10
Chronic Malnutrition	108	16.00	563	84.00

Table 3. Age distribution of chronic malnutrition among children registered at Sickle Cell Institute Chhattisgarh

Age Group in Years	Number	%
0-5	13	12.04
6-10	28	25.93
11-18	67	62.04
Total	**108**	**100**

Table 4. Risk factors among children with chronic malnutrition

Risk Factors	Number	%
Needed hospitalization		
Yes	**25**	**23.15***
No	**83**	**76.8**
Age group		
11-18 Years	**16**	**64***
Up to 11 Years	**9**	**36**
Blood type		
O Rh+	**43**	**39.81**
Other Blood groups (A+, AB+ and B+)	**65**	**60.18**

* p≤0.001

Table 5. Morbidity profile of children at the time of registrationat Sickle Cell Institute Chhattisgarh

Morbidity Pattern (N=671)	Number	%
Needed hospital care	180	26.8
Icterus and Pallor	186	27.7
GB pain and VOC	189	28.2
Asymptomatic	99	14.8
Bone and Joint Pain	17	2.5
Status of Anemia (N=447; status unknown: 224)		
No anemia	28	6.3
Mild	96	21.5
Moderate	215	48.1
Severe	108	24.2

Table 6. Nutritional status of children registeredat Sickle Cell Institute Chhattisgarh by whether or not hospitalization was needed

Nutritional Status	Hospitalization N (%)	No Hospitalization N (%)	Total
Underweight	104(27)	285(73)	389
Stunted	56(22)	197(88)	253
Wasting	76(29)	185(71)	261
Total	**236**	**667**	**903**

Table 7. Association of Adult Hemoglobin and Fetal Hemoglobin with Chronic malnutrition among children of Sickle Cell Institute Chhattisgarh

Chronic malnutrition	Hb level < 10 mg N(%)	HbF level < 20 N(%)	P value
Present	68(71.57)	27(28.43)	P = 0.005
Absent	40(33)	81(67)	
Total	108(100)	108(100)	

Hb = hemoglobin; HbF = fetal hemoglobin

Table 8. Association of underweight with age and sex and stunting with O Rh+ blood group among children of Sickle Cell Institute Chhattisgarh

Age group	Underweight N (%)		
	Male	Female	
Younger children (< 10 Yrs)	107(63.58)	64(37.42)	P = 0.005
Younger children (> 10 Yrs)	113(51.84)	105(48.16)	
Total	**220(56.56)**	**169(43.44)**	
	Stunting		
O Rh+ Blood group	Stunting	Normal	
Present	95(51.13)	63(39.87)	P = 0.005
Absent	158(37)	269(63)	
Total	**253(43.25)**	**332(56.75)**	

DISCUSSION

Adults and children with sickle cell anemia have a relative energy shortage. Hibbert et al. (2006) have shown that nutritional deficiency in SCA is secondary to a marked hyper-metabolic state associated with higher energy requirements. 16% of the children who have presented at the Sickle-cell Institute Chhattisgarh had chronic malnutrition and a high proportion had severe anemia, with Hb <10 mg/dl, with older children more likely to be malnourished compared to the younger ones (P < 0.01).

A similar study done by Boadu et al. (2018) observed that more than a third of the study children (38%) were malnourished, with stunting at 37.7% along with underweight and wasting at 58%, and 38.9% respectively. Again, their older children were more often malnourished. There is no difference in the rate of SCD by sex because it is not a sex-linked disease. In the present study, the SCD was more prevalent in males (56.6%) (p<0.05),whereas Masese et al., (2020) found the diseaseto be more prevalent in females (56%).

In this study, the prevalence of SCD was mainly found in people in the Other Backward Categories (72.11%) and 6.11% SCD were from the scheduled tribe categories. In a study conducted by Bhatia (1987), the prevalence of sickle cell carriers among different tribal groups varied from 1 to 40 per cent. (The Government of India identifies the most under-privileged groups as "scheduled caste" and "tribal" (indigenous groups). Those slightly less under-privileged are called "other backward categories".). We found a high prevalence of blood type O+ (39.8%). Similar results were found by Amodu et al. (2012), with 47.7%. Reduced dietary intake exists in SCA patients, and the resulting state of undernutrition and poor growth (Boadu et al. 2018) is due wholly or in part to reduced dietary and energy intake. In the present study, the prevalence of underweight, stunting and wasting were 5.8%, 37.7% and 38.9% respectively. However, a study of SCD patients in Cameroun by Charlotte et al. (2022), found a prevalence of underweight, stunting and wasting were only 3.6%,9.1% and 7.1% respectively.

It is becoming more apparent that dietary recommendations for SCD should include more emphasis on adequate amounts of macronutrients (Umeakunne and Hibbert, 2019). Singhal et al. (2002) measured the dietary intakes and resting metabolic rates (RMR) of 41 children with SCD and 31 control subjects, concluding that there is a relative

energy deficiency in SCD. The standard treatment protocol provides these supplements. One small study showed the efficacy of including macronutrient supplementation (Singh et al., 2023). We found chronic malnutrition to be significantly higher (39.81%) among O+ blood group in comparison to other blood groups ($p<0.05$). We found that at the time of diagnosis, the most common symptom was generalized bodyache and vaso occlusive crisis (28.17%). In a similar study by Singh et al. (2023), 37% among SCD patients were suffering crisis episodes.

In the current study, patients with Hb level > 10mg and HbF > 20% were significantly less at risk of hospitalization for complications of SCD; exactly the same observation were noted by Singh et al. (2023) among SCD patients with an HbF level> 20%. Evidence that nutrition intervention can reduce complication rates, length of hospital stay, readmission rates, mortality, and cost of care were documented by Tappenden et el. (2013).

CONCLUSION AND RECOMMENDATION

Our findings suggest that older children with the O+ blood group have increased risk of hospital admission and should thus be tracked on a priority basis. We found that in children presenting with SCD, a significant proportion had chronic malnutrition mixed with underweight, stunting and wasting. Stakeholders should work in the direction of developing a comprehensive nutrition care and education plan. Chronic malnutrition needs a combination of package of nutrition interventions. On this issue, there should be intense research in the field of nutrition specially focusing on how to increase red blood cells, as there is a high level of destruction of red blood cells and a decrease oxygen carrying capacity in SCD patients. Replenishment of red blood cells needs substrates such as protein and folic acid. SCD care and management is complex and multifactorial includes nutritional challenges at home as well as at hospital. Nutrition intervention will definitively improve standard care practice, increase survival and quality care of SCD children.

AUTHOR CONTRIBUTIONS

Author AS was involved in concept planning and data analysis. Author PKP contributed designing manuscript framework and author PSP made effort in manuscript writing.

CONFLICT OF INTEREST

The authors declare that they have no other potential conflicts of interest.

ACKNOWLEDGEMENTS

None

FUNDING

None.

Received: October 3, 2024; **Revised**: November 26, 2024; **Accepted**: December 9, 2024; **Published**: December 31, 2024.

REFERENCES

Amodu O.K., Olaniyan, S.A. Adeyemo, A.A. Troye-Blomberg, M. Olumese, P.E. & Omotade, O.O. 2012. Association of the sickle cell trait and the ABO blood group with clinical severity of malaria in southwest Nigeria. *Acta Tropica,* 123(2):72-77. https://doi.org/10.1016/j.actatropica.2012.03.013

Bhatia H.M., & Rao V.R. 1987. Genetic atlas of Indian Tribes. Bombay: Institute of Immunohaematology (ICMR). https://books.google.co.in/books/about/Genetic_Atlas_of_the_Indian_Tribes.html?id=ZtM4GwAACAAJ&redir_esc=y

Boadu, I., Ohemeng, A. & Renner, L.A. 2018. Dietary intakes and nutritional status of children with sickle cell disease at the Princess Marie Louise Hospital, Accra — a survey. *BMC Nutrition*, 4:33 https://doi.org/10.1186/s40795-018-0241-z

Charlotte E.E., Ritha Carole M.B., Calixthe I.P., Jeanne Georgette M.E., Patricia E., Iyawa H., Edgar M.M.L., Michael Ngenge B., Gaelle N.K., Claudia E.B.B., Dicka S.M., Paul K.N., & Daniele-Christiane K.M.K. 2022. Describing the growth and nutritional status of sickle cell disease children and adolescents with reference to WHO growth standards in Cameroon. *BMC Nutrition,* 8:154.

Eaton W.A., & Hofrichter J. 1987. Hemoglobin S gelation and sickle cell disease. *Blood*, 70(5):1245-1266.

Hibbert J.M., Creary M.S., Gee B.E., Buchanan I.D., Quarshie A., & Hsu L.L. 2006. Erythropoiesis and myocardial energy requirements contribute to the hypermetabolism of childhood sickle cell anemia. *Journal of Pediatric Gastroenterological Nutrition*. 43(5):680-687. doi: https://doi.org/10.1097/01.mpg.0000228120.44606.d6

Hyacinth H.I. 2018. Sickle-cell anaemia needs more food? *The Lancet Haematology,* 5(4):e130. https://doi.org/10.1016/S2352-3026(18)30032-2

Masese, R.V., Dominique B., Mitchell K., Liliana P., Eleanor S, Jane S. Hankins, M. T., Allison A.K., Victor R.G., Julie K., Robert G., Jeffrey A. G., Paula T., Nirmish S. 2020. Sex Based Differences in Sickle Cell Disease. *Blood,* 136(S1):37. https://doi.org/10.1182/blood-2020-140896.

https://doi.org/10.1182/blood-2020-140896

Ministry of Health and Family Welfare, Government of India. 2016. Prevention and control of hemoglobinopathies in India. https://nhm.gov.in/images/pdf/in-focus/NHM_Guidelines_on_Hemoglobinopathies_in_India.pdf

Ministry of Health and Family Welfare, Government of India. 2023. *Guidelines for National Programme for Prevention and Management of Sickle Cell Disease National Sickle Cell Anaemia Elimination Mission 2023.* https://sickle.nhm.gov.in/uploads/english/OperationalGuidelines.pdf

Platt O.S., Brambilla D.J., Rosse W.F., Milner P.F., Castro O., Steinberg M.H., & Klug P.P. 1994. Mortality in sickle cell disease. Life expectancy and risk factors for early death. *New England Journal of Medicine.* Jun 9;330(23):1639-1644. doi: https://doi.org/10.1056/NEJM199406093302303

Platt O.S. 2008. Hydroxyurea for the treatment of sickle cell anemia. *New England Journal of Medicine,* 358:1362–1369. https://doi.org/10.1056/NEJMct0708272

Singh A, Bokade C, Tirpude B, Suryawanshi MM, & Rohadkar LA. 2023. Clinical profiles of children with sickle cell anaemia presenting with acute clinical events: a single-center study. *Cureus*, 15(5):e39008. https://doi.org/10.7759/cureus.39008

Singhal A., Parker S., Linsell L., & Serjeant G. 2002. Energy intake and resting metabolic rate in preschool Jamaican children with homozygous sickle cell disease. *American Journal of Clinical Nutrition*. 75(6):1093-7. doi: https://doi.org/10.1093/ajcn/75.6.1093

Tappenden K.A., Quatrara B., Parkhurst M.L., Malone A.M., Fanjiang G., & Ziegler T.R. 2013. Critical role of nutrition in improving quality of care: an interdisciplinary call to action to address adult hospital malnutrition. *Journal of the Academy of Nutrition and Dietetics,* 113(9):1219-3 https://doi.org/10.1016/j.jand.2013.05.015

Umeakunne K., & Hibbert J.M. 2019. Nutrition in sickle cell disease: recent insights. *Nutrition and Dietary Supplements*, 23:9-17. https://doi.org/10.2147/NDS.S168257