

## DIETARY PREFERENCES AND PATTERN IN CHILDREN WITH SICKLE CELL DISEASE IN ENUGU

By

EZEPUE U. F. FMC<sub>Ophth</sub>, DCEH.<sup>1</sup> EMODI I. J. FMC<sub>Paed</sub><sup>2</sup>

OKAFOR U. FMC<sub>Paed</sub><sup>2</sup> OKONKWO P. O.<sup>3</sup> UWAKWEM A. C. FWACS<sup>1</sup>

<sup>1</sup>Department of Ophthalmology, UNTH Enugu <sup>2</sup>Department of Paediatrics, UNTH Enugu <sup>3</sup>Department of Pharmacology and Therapeutics, College of Medicine, UNN, Enugu Campus.

### SUMMARY

The looks of children with sickle cell anaemia give an impression of malnourishment. Not growing optimally, they have lower weight, height and muscle bulk in the upper limbs than age and sex matched controls. Their underlying state imposes an increased metabolic demand on them. Yet there is simultaneous inadequate nutritional intake and malabsorption in periods of crises. Evidence of depressed appetite also exists. Parents try to combat this through administration of blood tonics.

As a first step in assessing the nutritional status of children with sickle cell disease it was decided to conduct a detailed dietary interview. The food intake of children with sickle cell disease attending the sickle cell clinic of the University of Nigeria Teaching Hospital, Enugu was analyzed and compared with that of age matched controls that do not have sickle cell disease. Only differences which P-value was or 0.05 below were accepted as statistically significant.

In the age stratum 4 years and under, generally no significant differences were noted in the food consumption pattern of children with sickle cell disease when compared with age-matched controls. However, children with sickle cell disease consumed significantly more spinach than controls. In the age stratum 5-16 years, children with sickle cell disease consumed more of the food items considered.

Foods were sourced from mainly the market for all children and when not consumed, it was for similar reasons, mainly expensiveness and child's dislike. Night blindness seems to occur more in children with sickle cell disease.

It is concluded that parents do not purposely bias quantity or quality of food presentation to children with sickle cell disease despite noticing that relative to their other children, these children looked always ill-fed. The value of taking the specific nutritional needs of these children into considerations in food presentation was discussed. In addition, it is suggested that more attention be paid to nutrition and that nutritionist be involved in the health care and management of these children.

**Key Words:** Sickle Cell Disease; Dietary Preferences; Haemoglobinopathy in Enugu

### INTRODUCTION

The thinness and pallid complexion of children with sickle cell anaemia gives a general impression of a poor nutritional state. Optimal growth and development is dependent on adequate intake of a varied diet and good absorption of food offered. It has been documented that these children have significantly lower weight and height than age and sex matched controls<sup>1, 2</sup>. They also have a significantly lower muscle bulk in the upper limbs<sup>2, 3</sup>. It has also been documented that they have a higher metabolic rate secondary to both the increased tissue turn over in the bone marrow and the increased cardiac work<sup>4</sup>. The increased faecal nitrogen loss in these children has also been suggested as evidence of malabsorption<sup>5</sup>. More credence is given to this in

both children and adult sickle cell disease patients in whom it has been shown that an abnormality in D-Xylose absorption occurs<sup>6, 7</sup>.

It is thus obvious that the child with sickle cell anaemia needs extra nutritional intake to stay in positive calorie balance. Odunkor et al<sup>8</sup> in 1984 documented a decreased energy intake using a 24 hour dietary recall. Heyman et al<sup>9</sup> showed that the use of nasogastric tube feeding at night resulted in increased growth of two of their five patients who were small for age. This seems to be a way of bypassing the depressed appetite which might occur or increasing the energy intake of these children.

Parents bringing their children to our sickle cell disease clinics have been known to give the children blood tonics in order to build up the

### Correspondence Author

Dr. Ezepue U.F., Dept. of Ophthalmology,  
University of Nigeria Teaching Hospital, PMB 01129, Enugu.  
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children. This may be another means of increasing the child's appetite.

We are not aware of any study specifically comparing the dietary pattern of sickle cell disease patients with that of other persons who do not have sickle cell disease.

In view of this it was thought necessary to document and analyze the food intake preferences of these children and also find out whether the parents made any extra effort to vary this and any factors responsible for any shortcoming in food presentation to these children. Knowledge of this would enable better plans of management of these children's nutritional needs and education of parents.

**MATERIALS AND METHODS**

Consecutive known paediatric sickle cell patients attending the sickle cell clinic of the University of Nigeria Teaching Hospital (UNTH) Enugu, over a two month period were recruited for the study. The age limits were set at between 1 and 16 yrs. Only children completely off breast milk were recruited. There were no other exclusion

criteria except when the child or parent or guardian refused permission for inclusion in the study.

For control these children were compared with similar children who do not have sickle cell disease who attended the general paediatric and the eye clinic of the UNTH during the same period.

Personal data were collected and recorded in a previously prepared research questionnaire. These included age (rounded up or down to the nearest full age), sex and educational status of parents. In addition detailed dietary interview was conducted including consumption of common staples, common sources of animal protein, fruits and vegetables. Of interest were the frequency of consumption of the particular food item, source of the item and reason for non-consumption if not consumed.

The results of these were entered into the computer and analyzed with the EPI Info version 6 software. Differences between the cases and the controls were adjudged significant if the P-value were 0.05 or below.

**RESULTS**

Table 1 gives the Sex/Age distribution of the study population. The mean age of the 52 sickle cell disease (SSD) patients was 10.23yrs with a standard deviation (SD) of 4.40. The standard error of the mean (SEM) was 0.6102. The confidence interval was 9.004 to 11.456.

**TABLE 1  
SEX/AGE DISTRIBUTION OF THE STUDY POPULATION**

AGE GROUP	CHILDREN WITH SICKLE CELL DISEASE				CONTROLS			
	MALE	FEMALE	TOTAL	%	MALE	FEMALE	TOTAL	%
1 - 4	4	5	9	17.3	8	8	16	31.4
5 - 8	7	0	7	13.5	7	11	18	35.3
9 - 12	10	7	17	32.7	5	9	14	27.5
13 - 16	11	8	19	36.5	0	3	3	5.9
TOTAL	32	20	52	100%	20	31	51	100%

Among these patients, males had a mean age of 10.3 yrs with a SD of 4.692, SEM of 0.8294 and 95% confidence interval of 8.608 to 11.992.

Females had a mean age of 10.188 with a SD of 4.276, SEM 0.9561 and 95% Confidence

interval of 8.187 to 12.189.

No statistically significant difference was noted between the mean age of the males and females. The difference between the means is -0.1120 (95% confidence interval is -2.710 to 2.480).

T-test result at 50 degrees of freedom is 0.08658 and two tailed p-value is 0.9314.

For the controls, 51 in all, the mean age was 6.96 years with SD of 3.55 and SEM of 0.4971. The 95% confidence interval of the mean was 5.961 to 7.959. Males amongst the controls had a mean age of 6.050, SD of 3.84 and SEM of 0.8587. The 95% confidence interval (CI) was 4.253 to 7.847. The females had mean age of 7.548, SD of 3.641 and SEM of 0.6539. For females, the 95% CI was 6.213 to 8.883. Difference between the males and females was not found to be statistically significant the two sub-populations being homogenous. The actual difference between the mean ages was - 1.498 with 95% C.I. of 0.6457 to 3.642. The difference was tested with t-test which gave  $t=1.404$  at 49 degrees of freedom. The two tailed p-value of 0.1669 was adjudged not statistically significant.

Comparing the SSD patients and the controls, the mean ages differ significantly. The mean difference between the mean ages is 3.270 (1.706 to 4.834). Application of t-test at 101 degree of freedom gave  $t=4.146$ . A two tailed p-value of  $<0.0001$  is considered extremely significant. Although the difference between the two standard deviations were considered statistically not significant ( $F = 1.536$ , P-value = 0.0655) the Null-hypothesis of same mean ages was rejected.

Age is a known confounder in relation to food presentation and acceptance. Across the board comparison was therefore rejected. To control for the confounding effect of age, therefore, it was decided in the subsequent analysis, to stratify by age. Within the groups, it was noted that the food intake pattern differed between the children aged 1 to 4 years on the one hand and those aged 5 years and above on the other. Age stratification was based on this for all further analyses.

Tables 2 to 9 present the patterns of consumption of the various foods, the sources and reasons given for non-consumption. The pattern of consumption of each food item was compared between patients of sickle cell disease and normal children. The comparison was separate for each of two age group strata (1-4years and 5-16years). Only differences in consumption pattern whose probability of occurrence by chance (p-value) was at most 0.05 were accepted as being statistically significant.

Among the age stratum 1-4years (table 2 to 4) the food consumption pattern was similar, except for spinach consumption which sickle cell disease patients consumed more ( $X_2^2 = 4.95$ . Fisher's Exact 2 Tail P- value = 0.06).



**Table 4**  
**Pattern of Consumption of Select Green Leafy Vegetables (GLV) and Fruits by the Study Population aged 4 Years and Under.**

	% For Specific Food									
	% For Specific Food					% For Specific Food				
	Children with Sickle Cell Disease					Controls				
<b>Frequency of Consumption</b>	Mango	Pawpaw	Spinach	Other GLV	Carrots	Mango	Pawpaw	Spinach	Other GLV	Carrots
Never Taken			14.3		50.0	7.1	14.3	91.7	14.3	21.4
Several Times Daily				16.7		7.1			57.2	
About Once Daily		14.3	42.8	66.6	16.7	21.5	7.1		7.1	
At Least Once Weekly	28.6	42.8	28.6	16.7		14.3	42.9	8.3	14.3	21.4
At Least Once Monthly		14.3					7.1			
Only Occasionally	71.4	28.6	14.3		33.3	50.0	28.6		7.1	57.2
<b>Total</b>	100.0	100.0	100.0	100.0	100.0	100.0	100.0	100.0	100.0	100.0

Most differences in consumption pattern were noted in the age stratum 5 to 16 yrs, tables 5 to 7.

**Table 5**  
**Pattern of Consumption of Staple Foods by Study Population Aged 5 Years and above**

	% For Specific Food															
	Children With Sickle Cell Disease						Controls									
	Yam	Cassava	Garri	Rice	Beans	Cocoyam	Plantain	Potato	Yam	Cassava	Garri	Rice	Beans	Cocoyam	Plantain	Potato
<b>Frequency of Consumption</b>																
Never Taken		22.0	2.3		7.1	2.4	2.4			11.4				37.1		2.9
Several Times Daily			4.7						2.9							
About Once Daily	30.2	22.0	58.1	28.6	23.9	9.8	7.3	2.4	37.1	20.0	68.6	2.9	11.4			2.9
About Once Weekly	67.4	24.4	30.2	71.4	57.1	22.0	41.5	48.8	57.1	22.9	20.0	94.2	80.0	17.1	25.7	20.0
At Least Once Monthly	2.4	2.4			11.9	34.1	24.4	14.6	2.9	5.7		2.9		2.9	11.4	14.2
Only Occasionally		29.2	4.7			31.7	24.4	34.2	2.9	37.1	11.4		8.6	42.9	62.9	60.0
<b>Total</b>	100.0	100.0	100.0	100.0	100.0	100.0	100.0	100.0	100.0	100.0	100.0	100.0	100.0	100.0	100.0	100.0



Apart from cocoyam ( $X^2_2 = 8.96, P = 0.003$ ) and eggs ( $X^2_2 = 5041$  Fishers exact 2 tail  $P$  - value = 0.04) consumed more by control children, the children with sickle cell disease appeared to consume more of the items considered. These included plantain ( $X^2 4.26, P = 0.04$ ), potatoes ( $X^2 6.44 p < 0.01$ ), liver ( $X^2 = 4.48, p < 0.03$ ), multivitamin preparations ( $X^2 = 40.04, p < 0.000$ ) and pawpaw ( $X^2 = 5.67, p < 0.02$ ).

For the other food items, there was no statistically significant difference in consumption pattern between the control children and those with sickle cell disease.

Similarities were noted in the sourcing of, the food items for the families of these children the foods being mainly sourced from the market, table 8.

**Table 8**  
Sources of Food Items for Different Families

Source	Frequency % of Utilization of Source		
	Children With Sickle Cell Disease	Controls	Total
Farm Only	6.09	9.29	7.67
Market Only	83.10	76.08	79.63
Both Farm and Market	10.81	14.63	12.7
<b>Total</b>	<b>100.0</b>	<b>100</b>	<b>100.0</b>

When not consumed the reasons for non-consumption were also similar, mostly relative expensiveness and child's dislike particularly in the older child with sickle cell disease than in their age-matched controls. Prominent among the other unlisted reasons given for non consumption of particular food items is ignorance of nutritional value of item by the food providers, (table 9).

**Table 9**  
Reasons Given for Non-Consumption of Various Food Items

Reason	Frequency % of Utilization of Reason		
	Children With Sickle Cell Disease	Controls	Total
Not Available in Area	1.40	9.55	7.23
Too Expensive	18.31	17.42	17.67
Child Does Not Like it	21.13	16.85	18.07
Child Too Young to Take it	0	3.93	2.81
It is Bad for Children	0	1.69	1.21
Others*	59.16	50.56	53.01
<b>Total</b>	<b>100.0</b>	<b>100</b>	<b>100.0</b>

\*Others include ignorance of food providers on the

nutritional value of, or need to give children particular food item

Night blindness appeared to occur more in the children with sickle cell disease. The only 4 cases recorded occurred in children over 5 yrs of age with sickle cell disease and none in the control children, table 10.

( $X^2_2 = 3.45$ ). Fishers Exact 2 tail  $p$ -value = 0.12 which although marginally significant fails to attain our predetermined probability level.

**Table 10**  
History of Night Blindness

	NIGHT BLINDNESS		
	YES	NO	TOTAL
Children SSD	4	39	43
Control Children	0	35	35
<b>Total</b>	<b>100.0</b>	<b>100</b>	<b>100.0</b>

No parent was found to have purposely biased food presentation (neither in quantity nor in quality) to these sickle cell disease children compared to what is presented to other children in the household. However most parents noticed that these children still look ill-fed compared to their other children despite eating the same type of food.

## DISCUSSION

Sickle cell disease (SCD) is a problem of the black race. It is thus expected that research to improve its management would be the black races' problem and, indeed, it is. There are two major problems of the SCD patients: survival and quality of life. Attention to nutrition could improve these.

This study shows in tables 2 and 5 that the consumption pattern of major items of the diet do not differ in the younger paediatric SCD patients when compared with their peers who do not have SCD. This is not surprising because at this age group, parental influence play major roles in what the children take. Table 9 demonstrates that in children, reasons for not consuming any particular food item is mainly ignorance of food providers (parents/guardians). It is therefore obvious that the child's preferences, although noted, may be suppressed. Consequently the increased needs of these SCD children are hardly taken note of nor satisfied. Among the older children, child's preference becomes more pronounced. However, the differences are noted only in relatively uncommon

foods. The common foods are consumed in the same pattern across the age groups. Once again child's increased needs are not considered in the course of food presentation.

For all the families recruited for the study, the items assessed were sourced mostly from the market (table 8). This sourcing of these items from the market for both groups of patients means that the major determinant of the food any of the children actually takes are knowledge and the economic capabilities of the parents; and the child's preferences.

The role of family economic variables on nutrition becomes much more prominent in families with chronically ill members, such as sickle cell disease children. We believe that the frequent hospitalization and purchase of medication tend to drain the family resources. Frequently, there are little funds left after these expenses to spend on quality food for the family. This shows easily in the children primarily because of their increased needs. The even greater needs of the children with sickle cell disease explain their worse nutritional outlook despite eating the same food as other children in the family.

We noticed that child's preference appears more important in the SCD patients than in the others when a particular food is not consumed and it is easier to notice in the older children. This may be a reflection of the effect of SCD on the appetite. There is therefore a need to improve their appetite through more persuasion or some form of appetite stimulation. Occasionally, different, novel and imaginative methods of food preparation and presentation could help improve consumption. This is very important because although it has been shown that nasogastric tube feeding improve outlook and weight gain<sup>9</sup>, it is not a practical method in all cases.

We found that parents are generally unaware that SCD patients have special nutritional needs. Specifically, due to their high metabolic rate consequent on normal growth for their age added to their increased bone marrow cell turnover, they need increased quantities of qualitative food relative to their normal age mates. This need cuts across all food types - both macronutrients, minerals and micronutrients for reasons already stated earlier.

It has been shown that there is increased production of singlet oxygen<sup>10</sup>, hydrogen peroxide<sup>11</sup>, and the hydroxyl ion<sup>10</sup> by the sickle cell haemoglobin (HBSS). The availability of free iron in the tissues and blood catalysis the production of these superoxides<sup>11</sup>. It is now well known that these

free radicals damage cell lipid membrane. Thus in SCD patients this damage adds to the premature ageing and haemolysis of the red blood cells and anaemia<sup>12,13</sup>. Stone et al<sup>14</sup> in their review suggest that anti-oxidants could decrease severity of disease in these patients, and thereby influence morbidity and quality of life. Indirectly too, survival would also be affected.

Antioxidants can be supplied by both medication, and most importantly, diet. The carotenes of the diet are well known antioxidants. They are supplied by diets containing yellow fruits, dark green leafy vegetables and others found abundantly in this part of the world. Preformed vitamin A and zinc found in other dietary sources are also effective in mopping up free radicals.

Because of the foregone, we studied the prevalence of nightblindness in the study population. Nightblindness is one of the early signs of vitamin A deficiency. We believed that the children with sickle cell disease the production of free radicals by the frequent haemolysis of red cells and the mopping up of these by antioxidants would put stress on the supply of these antioxidants. These children would consequently be at increased risk of vitamin A deficiency and other antioxidants. Our tests of this hypothesis gave equivocal results and studies of larger populations would be required to prove or disprove this.

Parents and carers of SCD patients need to be educated by physicians and other health workers on the increased nutritional needs of these children and, of course, older patients. This single intervention would go a long way to improve on the perpetual pallid appearance of these patients. There is also a need for micronutrient supplementation in these patients because of their increased nutritional needs.

Equally important is the need to get specialists in nutrition and dietetics involved in the management of SCD patients. Hitherto, it has been the duty of only physicians with little expert knowledge in nutrition to manage these patients.

In addition, nutrition experts should be called upon to get involved in research aimed at improving the consumption of healthy food by these patients. The preparation of high-energy-density porridge by malting of a mixture of corn plus soya beans and or cow peas can increase the caloric and protein content without increasing the volume.<sup>15</sup>

There is still need for more research aimed at understanding and managing the malabsorption



present in these children. Otherwise, any improvement achieved in appetite stimulation and food presentation could still be brought to naught by poor absorption. In addition, operation research to test the usefulness of these suggestions should be conducted.

### CONCLUSION

Often due to ignorance, parents and food providers in families with sickle cell disease children do not make any special effort to take care of the special nutritional needs of these children through well varied diets.

Empowering these parents and food providers economically and particularly through the provision of appropriate nutrition education could help improve the nutritional outlook of children with sickle cell disease.

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