

## Assessment of the Nutritional Status & the Dietary Pattern of the Under 5 Years Old Children with Sickle Cell Disease in Basrah Center for Hereditary Blood Diseases

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### ABSTRACT:

#### BACKGROUND:

Children with sickle cell disease (SCD) were more liable to growth impairment as compared with healthy children as result of increased energy demand, endocrine and inadequate dietary intake

#### OBJECTIVE:

To assess nutritional status, dietary pattern of children with SCD and to determine the relationship between sociodemographic characteristics & nutritional status of the children.

#### PATIENTS AND METHODS:

A cross sectional study conducted in Basrah center for hereditary blood diseases to evaluate the nutritional status of 60 patients all under 5 years with SCD by anthropometrics measurement, use of WHO growth charts, prestructured questionnaire and assess their dietary intake depending on recommendation of the integrated management of childhood illness.

#### RESULTS:

The study involved 60 children less than 5 years old with SCD, their mean age  $41 \pm 14$  months, 65% of them had (SS) genotype. 65% of them were suffering from moderate anaemia. The frequency of underweight, stunting, wasting and obesity were 6.7%, 10%, 1.7, and 1.7% respectively. There was no significant association of growth deficit in patient with SCD and following: gender, genotype, number of painful crisis, number of Blood transfusion in last year, mother education and dietary intake.

#### CONCLUSION:

There was low frequency of malnutrition in children with SCD in Basra. Growth monitoring and nutritional counseling are needed.

**KEYWORDS:** Sickle cell disease, Nutritional status, Dietary intake, Anaemia.

### INTRODUCTION:

Sickle cell disease (SCD) is one of the most common life-threatening hereditary diseases in humans and primarily affects people of African, Indian, and Arab ancestry<sup>(1)</sup>. Globally about 300,000 children are born with sickle cell disease every year<sup>(2)</sup>. The prevalence for sickle-cell trait was about 21% and for SCD was 2.6% by newborn screening for SCD in the Eastern province of Saudi Arabia over a 9-year period compared with 17% and 1.2%, respectively, from premarital screening<sup>(3)</sup>. In Iraq, the prevalence of SCD was 13.9 / 100,000 in 2015 and highest prevalence of SCD was recorded in Basra Province (124 / 100,000)<sup>(4)</sup>.

Sickle haemoglobin (HbS) inherited as an autosomal recessive. The most common and severe form is homozygous HbSS (sickle cell anaemia), other types of sickle cell disease include heterozygous conditions, such as haemoglobin C (HbC) with HbS (HbSC), HbS with  $\beta$ -thalassaemia (HbS/ $\beta$ 0-thalassaemia or HbS/ $\beta$ + -thalassaemia), HbSD and sickle cell trait (HbAS)<sup>(5)</sup>.

In 2019, there were 144 million of less than 5 years old children suffer from stunting, 47 million of them were wasted of which 14.3 million were severely wasted worldwide<sup>(6)</sup>. About (45%) of child deaths due to malnutrition, especially in low- and middle-income countries<sup>(7)</sup>. Approximately 41% of American children with SCD were moderately underweight and 25% of them were severely underweight and 11% of them were wasted<sup>(8)</sup>.

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SCD affects physical growth throughout childhood and early adolescence, and children with this condition are often leaner and shorter than their healthy peers <sup>(8,9)</sup>. The growth failure mechanism in SCD includes: interleukin-6-related appetite suppression resulted in decreased dietary intake, endocrine dysfunction, micronutrient deficiency, increased cardiac energy demand, increased red cell turnover, and hypermetabolism <sup>(9,10,11)</sup>. Patients with sickle cell disease had increased basal metabolic rate so they require much higher energy and protein consumption than healthy individuals and liable to suffer from undernutrition if energy intake is consistently low <sup>(12)</sup>.

It has been found that wasting is correlated with increased hospitalization and poor clinical outcomes in SCD <sup>(11,13)</sup>, also the World Health Organization (WHO) recognized that both overweight and malnutrition were associated with poor prognoses <sup>(2)</sup>. It has been recommended that Providing adequate nutrition to under five years children with SCD may improve their body composition, especially the lean body mass, and therefore will reduce the SCD morbidity and mortality <sup>(8,11,13)</sup>.

Growth monitoring is an important tool to assess the disease severity, the efficacy of medical and nutritional interventions <sup>(9)</sup>.

The current study was conducted to assess nutritional status, dietary pattern of children with SCD and to determine the relationship between sociodemographic characteristics & nutritional status of the under 5 years children with SCD in Basra.

#### **PATIENTS AND METHODS:**

A cross sectional descriptive study was carried out in Basra center for hereditary blood diseases (from 1st of February to the 30th of April 2021). The study included a total of 60 patients all under 5 years with SCD attending the center who did not present with painful crises, the study excluded all those with another chronic condition that may interfere with growth (chronic kidney disease, heart disease, endocrine disease). Clinical data were collected by direct interview according to a pre-structured questionnaire, hemoglobin level was measured for assessing anaemia severity then Hemoglobin level classified into 4 groups <sup>(14)</sup>:

1. gm/dl <7 1. Severe anemia

2. moderate 7 -9.9 gm/dl.

10-10.9gm/dl. 3. Mild

g/dl. ≥ 11.4. Normal

Disease severity was assessed according to the frequency of painful crises or blood

transfusion in last year. Patient with SCA is considered as having severe disease when he has ≥ 3 admission for vaso-occlusive crisis (VOC) last year or if he need ≥ 3 blood transfusions last year <sup>(15)</sup>.

Height and weight were measured to the nearest 0.1cm and 0.1kg, and Body Mass Index (BMI) was calculated by dividing weight (in kg) by the square of height (in meter):  $W/H^2$ , then the Z-score of these indicators were calculated: body mass index for age, weight for height, weight for age (W/A), and height for age, using Anthro 3.2.2 (WHO—Department of Nutrition for Health and Development, Geneva, Switzerland). Stunting, underweight, and wasting were defined by Z-score <-2 for height-for-age (HAZ), weight-for-age (WAZ) and BMI-for-age (BMI-Z), respectively. Stunting was considered mild for a Z-score between -1 and -2; moderate for a Z-score between -2 and -3 and severe for a Z-score <-3. Overweight and obesity were defined by BMI-Z above +2 and above +3 Z-score respectively <sup>(16)</sup>.

The dietary intake is considered adequate when it meets the feeding recommendations of the integrated management of childhood illness <sup>17</sup>.

Agreements of the Ministry of Health and research committee of Basrah Health Directorate were obtained before starting the study. Informed parental consent Obtained.

#### **Statistical analysis**

Data were analyzed using SPSS for Window version 25, Data were presented as frequencies, mean Fisher's exact tests were used to assess the correlation between the independent variables (age, sex, Diet intake, genotype, mother education & factors related to the severity of the disease), and the dependent variables (weight, height and BMI for age). All statistical tests were two-tailed with  $p < 0.05$  considered significant.

#### **RESULTS:**

The study involved 60 SCD children, their mean age  $41 \pm 14$  months, The general characteristics of the participating children are described in Table 1. 53.3% were males, 51.7% of participants were diagnosed with SCD during their first year of life. 63.3% were lived in rural area, 56.7% of them had no other sibling affected by hemoglobinopathy.

Sickle cell anaemia (SS) represents 65%, while Sickle thalassemia (S/B) form 28.3% of patients as shown in table 2. Mean haemoglobin of  $9 \pm 1.3$  g/dl. 65% of them were suffering from moderate anaemia as seen in table 3. Around three quarters 73.3% of patients had no

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admission for painful crisis and 50% of patients had no Blood transfusion last year.

The frequency of underweight, stunting and wasting were 6.7%,10% and 1.7% respectively as demonstrated in table 5.

There was no significant association of growth deficit in patients with SCD and following:

gender, number of painful crisis, number of Blood transfusion in last year, Mother Education, Dietary intake, genotype and Hb level. As seen in table 6.

**Table 1: Distribution of studied children according to Sociodemographic characteristics.**

| variables                                             |                      | No=60 | %    |
|-------------------------------------------------------|----------------------|-------|------|
| Age in months<br>(Mean =41±13.8)                      | <12                  | 2     | 3.3  |
|                                                       | 12-24                | 4     | 6.7  |
|                                                       | 24-36                | 14    | 23.3 |
|                                                       | 36-48                | 15    | 25   |
|                                                       | 48-60                | 25    | 41.7 |
| Gender                                                | Male                 | 32    | 53.3 |
|                                                       | Female               | 28    | 46.7 |
| Age of diagnosis in months                            | <12                  | 31    | 51.7 |
|                                                       | 12-24                | 16    | 26.7 |
|                                                       | 24-36                | 4     | 6.6  |
|                                                       | 36-48                | 5     | 8.3  |
|                                                       | 48-60                | 4     | 6.6  |
| Birth order                                           | First                | 15    | 25   |
|                                                       | 2-4                  | 40    | 66.7 |
|                                                       | ≥5                   | 5     | 8.3  |
| Residence                                             | Urban                | 22    | 36.7 |
|                                                       | Rural                | 38    | 63.3 |
| Mother Education                                      | Illiterate           | 3     | 5    |
|                                                       | Primary school       | 29    | 48.3 |
|                                                       | Secondary school     | 22    | 36.7 |
|                                                       | Higher Education     | 6     | 10   |
| Family size                                           | ≤ 3                  | 13    | 21.7 |
|                                                       | 4-6                  | 38    | 63.3 |
|                                                       | ≥ 7                  | 9     | 15   |
| Number of other sibling affected by heamoglobinopathy | No sibling affected  | 34    | 56.7 |
|                                                       | one sibling affected | 16    | 26.7 |
|                                                       | 2                    | 9     | 15   |
|                                                       | 4                    | 1     | 1.7  |

**Table 2: Distribution of sickle cell children according to Hb variant.**

| Type of Hb variant       | Number | %    |
|--------------------------|--------|------|
| Sickle cell anaemia (SS) | 39     | 65   |
| Sickle trait (AS)        | 2      | 3.3  |
| Sickle thalassemia (S/B) | 17     | 28.3 |
| Sickle (S/D)             | 2      | 3.3  |
| Total                    | 60     | 100  |

**Table 3: Distribution of sickle cell children according to hemoglobin level.**

| Hb gm/dl<br>(mean=9±1.3) | no | %   |
|--------------------------|----|-----|
| <7                       | 4  | 6.7 |
| 7-9.9                    | 39 | 65  |
| 10.1-10.9                | 12 | 20  |
| ≥ 11                     | 5  | 8.3 |
| Total                    | 60 | 100 |

**Table 4: Distribution of sickle cell children according to severity of disease by number of admission for vaso-occlusive crisis & number of blood transfusions received in the last year.**

| Clinical parameters         |                   | Number | %    |
|-----------------------------|-------------------|--------|------|
| VOC in last year            | no Admission      | 44     | 73.3 |
|                             | <3 painful crisis | 9      | 15   |
|                             | ≥3 painful crisis | 7      | 11.7 |
| Blood transfusion last year | No transfusion    | 30     | 50   |
|                             | <3 transfusion    | 12     | 20   |
|                             | ≥3 transfusion    | 18     | 30   |

**Table 5: Distribution of sickle cell children according to nutritional status & dietary pattern.**

| Variables         |                          | No=60 | %    |
|-------------------|--------------------------|-------|------|
| Weight for age    | Normal weight            | 55    | 91.7 |
|                   | Underweight              | 4     | 6.7  |
|                   | Over weight              | 1     | 1.7  |
| height for age    | Normal height            | 53    | 88.3 |
|                   | Stunting                 | 6     | 10   |
|                   | sever Stunting           | 1     | 1.7  |
| weight for height | Normal weight for height | 56    | 93.3 |
|                   | Wasting                  | 2     | 3.3  |
|                   | Overweight               | 1     | 1.7  |
|                   | Obesity                  | 1     | 1.7  |
| BMI               | Normal                   | 57    | 95   |
|                   | Wasting                  | 1     | 1.7  |
|                   | Overweight               | 1     | 1.7  |
|                   | Obesity                  | 1     | 1.7  |
| Diet intake       | Adequate                 | 47    | 78.3 |
|                   | Inadequate               | 13    | 21.7 |

Table 6: P-value by Fisher's Exact Test for factors associated with growth deficit in patients with SCA.

| variables                      |                          | Underweight<br>no=4 | Stunting<br>no=6 | Wasting<br>no=1 |
|--------------------------------|--------------------------|---------------------|------------------|-----------------|
| Gender                         | Male                     | 2 (3.3)             | 5(8.3)           | 0               |
|                                | Female                   | 2 (3.3)             | 1(1.7)           | 1(1.7)          |
|                                | P-value                  | 0.794               | 0.21             | 0.855           |
| VOC in last year               | no Admission for VOC     | 3(5)                | 5(8.3)           | 1(1.7)          |
|                                | 3< painful crisis        | 1(1.7)              | 1(1.7)           | 0               |
|                                | 3≥ painful crisis        | 0                   | 0                | 0               |
|                                | P-value                  | 0.801               | 0.321            | 1               |
| Blood transfusion in last year | No blood transfusion     | 4(6.7)              | 4(6.7)           | 1(1.7)          |
|                                | <3 transfusion           | 0                   | 1(1.7)           | 0               |
|                                | ≥3 transfusion           | 0                   | 1(1.7)           | 0               |
|                                | P-value                  | 0.283               | 0.926            | 1               |
| Mother Education               | Illiterate               | 0                   | 0                | 0               |
|                                | Primary school           | 2(3.3)              | 3(5)             | 0               |
|                                | Secondary school         | 2(3.3)              | 2(3.3)           | 1(1.7)          |
|                                | Higher Education         | 0                   | 1(1.7)           | 0               |
|                                | P-value                  | 0.389               | 0.895            | 0.436           |
| Diet intake                    | Adequate                 | 3(5)                | 6(10)            | 1(1.7)          |
|                                | In adequate              | 1(1.7)              | 0                | 0               |
|                                | P-value                  | 1                   | 0.085            | 1               |
| Type of Hb variant             | Sickle cell anaemia (SS) | 4(6.7)              | 5(8.3)           | 1(1.7)          |
|                                | Sickle trait (AS)        | 0                   | 0                | 0               |
|                                | Sickle thalassemia (S/B) | 0                   | 1(1.7)           | 0               |
|                                | Sickle (S/D)             | 0                   | 0                | 0               |
|                                | P-value                  | 0.659               | 0.855            | 0.190           |
| Hb level                       | Hb                       | 4(6.7)              | 6(10)            | 1(1.7)          |
|                                | P-value                  | 0.712               | 0.713            | 0.240           |

**DISCUSSION:**

The current study revealed that children with SCD usually were diagnosed earlier than Al-Saqladi AW et al study in Yemen, Boadu I et al study in Accra and Ngo Um SS et al study in Cameroon <sup>(9,18,19)</sup>, indicated either disease severity or voluntary screening by family who had other affected sibling, since there were no routine neonatal screening. Most of children had (SS) genotype similar to another studies <sup>(9,18)</sup>, reflecting high incidence of SCD in south of Iraq <sup>(4)</sup>. most of the patient were anemic as in the studies <sup>(9,18)</sup> while mean Hb level was higher than Islam MR et al study in Nigeria demographic and health Survey, Fadhil RS et al study in Basra, Ukoha OM et al study in southeast Nigeria <sup>(13,20,21)</sup> and also higher than other studies <sup>(9,18,19)</sup>, as result of proper management and adequate dietary intake. Around half of the patients were from rural area similar to the finding of studies <sup>(13,20)</sup>, since there were high consanguineous marriages in rural area. Most of patient had mild disease while in studies <sup>(9,20)</sup> most of them had severe disease.

The frequency of underweight, stunting and wasting were nearly similar to studies <sup>(19,21)</sup> but lower than Botelho EC et al study in Rio de Janeiro <sup>(22)</sup> and other studies <sup>(9,13,18,20)</sup>. Like children with other chronic diseases, children with SCD are at risk of growth deficit <sup>(23)</sup>, as result of anorexia leading to decreased dietary intake and increased basal metabolic rate due to protein hypermetabolism, increased cardiac energy demand, and increased red cell turnover <sup>(10)</sup>. The frequency of overweight and obesity was near to studies <sup>(20,21)</sup> but lower than Botelho EC et al study <sup>(22)</sup>, maybe due to less severity of illness, chronic transfusion, the use of hydroxyurea, increase consumption of ultraprocessed foods and urbanization <sup>(21,22,9)</sup>. There was no significant association of stunting and underweight with sickling genotypes while it was significant in Nigeria demographic and health Survey <sup>(13)</sup>, possibly due to small sample size. There was no significant association between disease severity and growth deficit as in Al-Saqladi AW et al study <sup>(9)</sup>.

Gender was not significantly associated with growth impairment similar to these studies <sup>(9,13)</sup>, as there was no great difference in nutritional demand between boys and girls at this age group. Although this study shows higher percentage of adequate dietary intake as compared with Boadu I et al study <sup>(18)</sup>, it was not significantly associated with growth indicators, Children with sickle cell disease require more energy and protein than healthy people, and are liable to be malnourished if their energy intake continues to be low <sup>(12)</sup>.

### CONCLUSION:

This study shows that children with SCD have low frequency of malnutrition in addition to overweight and obesity, most of them had adequate dietary intake, there was no significant association of growth indicators with gender, disease severity and mother education. Regular nutritional assessment of children with SCA together with nutrition education and counseling is needed.

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