Sickle cell anemia (SCA) is an inherited blood disorder. It is transmitted as an autosomal recessive gene characterized by recurrent chronic hemolytic anemia, vaso-occlusive crises, and predisposition to infections that seriously impact morbidity and mortality. SCA is common in Sub-Saharan Africa, the Caribbean, Middle East, and India, and the prevalence is increasing globally because of migration. The first reporting of the presence of the hemoglobin S (HbS) gene in Sudan was by Archibald. Since, different studies have revealed that Sudan has a high prevalence of SCA, with HbS allele frequency that ranges between 0.8% in the Northern regions and over 30% in the Western regions of the country. The high HbS allele frequency in the latter region is thought to be due to the high level of consanguineous marriages (40–45%), the tribal influx from West Africa, and endemic malaria.

Despite its genetic simplicity, where SCA is a single substitution base change T>A at codon 6 of the β gene, there is significant clinical heterogeneity of the disease probably attributable to genetic, epigenetic, nutritional, and many environmental factors interacting with each other. Generally, the care for patients with SCA has improved over the years due to earlier diagnosis, the widespread use of penicillin prophylaxis, vaccination, folate

**ABSTRACT**

Objectives: We sought to investigate the nutritional and hematological status of Sudanese women of childbearing age with sickle cell anemia (SCA). Anthropometry and hematology were used to assess nutritional status and health and disease conditions, respectively. Methods: Women with steady-state (HbSS, n = 39; age = 19.0±2.7) and without (HbAA, n = 36; age, 19.8±2.7) SCA were recruited during a routine visit to the Hematology Clinic, Ibn-Auf Teaching Hospital, Khartoum, Sudan. Results: The two groups of women lived in similar environmental conditions and ate similar diets three times a day. However, despite taking regular meals, the women with sickle anemia were thinner and lighter (p < 0.001) and shorter (p = 0.002) compared with those who do not have the disease. Also, they had higher levels of mean corpuscular hemoglobin (Hb) concentration and white cell count (p < 0.001), mean corpuscular volume (p = 0.003), and platelet (p = 0.002) and lower packed cell volume and Hb (p < 0.001). There was no difference in levels of anthropometric and hematological variables between the hydroxyurea treated and untreated SCA patients (p > 0.050). Conclusions: The low anthropometric (height, weight, and body mass index) and abnormal hematological values in the women with SCA in steady-state reflect sustained nutritional insults inflicted by the disease and poverty. Tailored nutritional counseling/advice must be an integral part of managing patients with SCA. Such advice is particularly vital for women of childbearing age because of the adverse effects of prepregnancy nutritional deficiency on birth outcomes.
supplementation, and access to comprehensive care programs including blood transfusions and hydroxyurea therapy, impacting significantly on morbidity and mortality.\textsuperscript{10–14}

Nutritional status and growth and development are important indicators of the overall health and well-being of patients with SCA. Indeed, patients with sub-optimal nutrition status are at increased risk of hospital admission and severe morbidity and mortality. These health indicators are often compromised/impaired by a high metabolic rate caused by the disease, reduced absorption of essential nutrients, and appetite loss induced by the disease and its treatments. Also, a lack of access to nutrient-dense foods is a major problem for sickle cell patients in economically disadvantaged countries and communities.

It is widely recognised that maternal nutritional status before pregnancy is one of the main determinate factors of pregnancy outcome.\textsuperscript{15–20} Published data are scarce on the nutritional status of women of childbearing age with SCA. Indeed, the effect of nutritional status before pregnancy on maternal and fetal outcomes has not been studied in women with the disease.

Until recently, it was a rarity to find pregnant women with sickle cell disease (SCD) in Sudan. However, because of improved disease management and the resulting increase in life expectancy, their number has increased significantly. Regardless, the country is highly underdeveloped with a rudimentary health service system and rampant poverty and malnutrition. In this highly patriarchal society, women, young children, and individuals with chronic diseases tend to bear the brunt of gross malnutrition and undernutrition.

The aim was to investigate the nutritional status and health of Sudanese women of childbearing age with SCA. Anthropometry and hematology were used to assess nutritional status and health and disease conditions, respectively.

**METHODS**

Women with steady-state SCA HbSS (n = 39) and without HbAA (n = 36) were recruited during a routine visit to the Hematology Clinic, University of Khartoum Ibn Auf Teaching Hospital. Unmarried women aged 16–40 years who volunteered to participate in the study and with the mental competence to give informed consent were included in the study. Women with sickle cell crisis, acute illness or blood transfusion in the previous four months, the presence of other chronic diseases or a physical disability that impairs access to food or restricts eating, and those that were pregnant were excluded from the study. Steady-state is defined absence of sickle cell crisis or acute illness for four months before and up to two weeks after blood collection for the study. A blood specimen (5 mL), detailed anthropometric, demographic data, medical history, and dietary habits were collected. We obtained ethical approval from the Ministry of Health of Sudan, University of Khartoum, Medical School, and London Metropolitan University, and signed informed consent from the participants.

Demographic data from the patients and medical history from hospital records and patients were collected using a questionnaire developed for the study.

Weight in kilograms and height in centimeters were assessed with a Seca Electronic Scale 890 (UNISCALE, Seca, Birmingham, UK) and a height-length measuring board (Schorr, Weight and Measure, LLC, Olney, Maryland, USA), respectively.

Hb concentration, packed cell volume (PCV), mean corpuscular volume (MCV), mean corpuscular hemoglobin (MCH), white blood cell (WBC) count and platelets count (PLTs) were measured using the Sysmex KX-21 N Automated Hematology Analyzer (Sysmex Corporation, Kobe, Japan).

Data are expressed as mean±standard deviation or percentages. Quantitative data were tested for normality and homogeneity of variance and subsequently analyzed with the independent $t$-test (parametric data) or the Mann-Whitney $U$ test (non-parametric data). Categorical (sociodemographic, clinical, and laboratory characteristics) data with cell frequency of five or more were assessed with a chi-square test on the contingency platform. When the observed cell count was less than five, chi-square and Yate’s correction of continuity and Fisher’s exact test were used under the assumption of independence of rows and columns and conditional on the marginal totals. The significance level was set at $p < 0.050$. The data was analyzed with SPSS Statistics (IBM Corp. Released 2019. IBM SPSS Statistics for Windows, Version 26.0. Armonk, NY: IBM Corp.).
RESULTS

The anthropometric and demographics characteristics of the women with (HbSS) and without (HbAA) SCA are presented in Table 1. There was no difference in age between the HbSS and HbAA groups ($p > 0.05$). However, the latter group was taller ($p = 0.002$), heavier ($p = 0.001$), and had a higher body mass index (BMI) compared with the former. The HbAA women had higher literacy and attendance of tertiary education rates ($p < 0.001$).

Most of the sickle cell patients and healthy control subjects eat regular breakfast (94.9 vs. 94.4%) and lunch (92.3 vs. 94.4%) [Table 2]. However, a significant number of the two groups do not consume dinner regularly (HbSS = 41.0% and HbAA = 61.1%, $p < 0.05$).

Of the HbSS, four had liver enlargement (4 cm, $n = 2$; 2 cm, $n = 2$) and one had spleen enlargement (6 cm). None of the patients had a scar for splenectomy. A few of the HbSS patients had a history of painful crisis with hemolysis and infection.

The women with SCA compared with the healthy control had lower PCV and Hb concentration ($p < 0.001$) and higher MCH ($p < 0.001$), MCV ($p = 0.003$), WBC ($p < 0.001$), and PLTs ($p = 0.002$) counts [Table 3]. There was no difference in lymphocyte and neutrophil counts and systolic and diastolic blood pressure between the two groups ($p > 0.05$).

Of the thirty-nine patients who consented to participate in the study, 25 were on hydroxyurea treatment. Anthropometric, demographic, and hematological variable data of the hydroxyurea treated and untreated patients at steady-state are presented in Table 4. There was no difference in age, weight, height, BMI, and systolic and diastolic blood pressure between the two groups ($p > 0.05$).

The hydroxyurea treated and untreated patients had a similar history of blood transfusion rates and comparable levels of Hb concentration, PCV, MCH, MCV, WBC, PLTs, neutrophil, and lymphocyte counts ($p > 0.05$). Also, the percentages of patients who were receiving folic acid supplementation were not different (treated 100% and untreated 92.8%, $p > 0.05$).

DISCUSSION

Consanguineous, marriage between first and second cousins, is a common tradition in all Sudanese tribes. This is evident from the high percentage of parental blood relationship of the HbSS (64.1%) and HbAA (44.4%) groups. Consistent with our findings, Daak et al\textsuperscript{5} reported high parental and self-consanguineous

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**Table 1:** Age, clinical, and demographic parameters of the participants.

| Characteristics                  | HbSS (n = 39) | HbAA (n = 36) | p-value |
|----------------------------------|---------------|---------------|---------|
| Age, years                       | 19.0 ± 2.7    | 19.8 ± 2.7    | NS      |
| Weight, kg                       | 39.0 ± 8.9    | 53.8 ± 11.0   | 0.001   |
| Height, cm                       | 151.0 ± 11.7  | 158.0 ± 6.7   | 0.002   |
| BMI, kg/m$^2$                    | 17.2 ± 4.3    | 21.5 ± 4.2    | 0.001   |
| Underweight (<18.5)              | 27 (69.2)     | 9 (25.0)      | 0.001   |
| Normal weight (18.5–24.9)        | 11 (28.2)     | 20 (55.6)     | < 0.001 |
| Overweight/obese (≥25.0)         | 1 (2.6)       | 7 (19.4)      | 0.209   |
| Education                        |               |               |         |
| Illiterate/primary school        | 24 (61.5)     | 3 (8.3)       | < 0.001 |
| Middle/high school               | 13 (33.3)     | 11 (30.6)     |         |
| University                       | 2 (5.1)       | 22 (61.1)     | < 0.001 |
| Parental relationship            |               |               |         |
| First- and second-degree relatives | 25 (64.1)   | 16 (44.4)     | 0.037   |
| Unrelated                        | 13 (33.3)     | 20 (55.6)     | 0.001   |
| Siblings with sickle cell disease|               |               |         |
| Yes                              | 22 (56.4)     | 3 (8.3)       | < 0.001 |
| No                               | 17 (43.6)     | 33 (91.7)     | < 0.001 |

NS: not significant; BMI: body mass index.

**Table 2:** Daily food consumption frequency of the participants.

| Response           | HbSS (n = 39) | HbAA (n = 36) | p-value |
|--------------------|---------------|---------------|---------|
| Regular breakfast  |               |               |         |
| Yes                | 37 (94.9)     | 34 (94.4)     | NS      |
| No                 | 2 (5.1)       | 2 (5.6)       |         |
| Regular lunch      |               |               |         |
| Yes                | 36 (92.3)     | 34 (94.4)     | NS      |
| No                 | 3 (7.7)       | 2 (5.6)       |         |
| Regular dinner     |               |               |         |
| Yes                | 23 (59.0)     | 14 (38.9)     | NS      |
| No                 | 16 (41.0)     | 22 (61.1)     |         |

NS not significant.
marriages in patients with SCD in Western Kordofan State, Sudan. In this study, although a significant number of the control and patients were born to parents who are blood relatives, the figure was more remarkable in the SCD group. It may be that the social stigma associated with the disease makes it very difficult for affected individuals to find a suitor (husband/wife) outside the immediate family circle. Regardless, as it is apparent from the number of siblings with SCA in the patient group, consanguinity/in-breeding would be expected to perpetuate and increase the prevalence of the disease in Sudan.

There is an urgent need for comprehensive and well-planned education, social awareness, and counseling programs targeted for patients with SCA, and the community, particularly in rural areas with deep-rooted traditions, to help reduce discrimination and stigma. In Sudan, the strong predictors of negative attitude and discrimination against SCD are lacking knowledge and low socioeconomic status. Similar observations have been reported from other Sub-Sahara African countries.

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### Table 3: Hematological characteristics of the participants.

| Characteristics | HbSS (n = 39) | HbAA (n = 36) | p-value |
|-----------------|---------------|---------------|---------|
| Hb, g/dL        | 9.2 ± 1.6     | 14.3 ± 1.6    | < 0.001 |
| PCV, %          | 25.1 ± 6.2    | 37.6 ± 3.3    | < 0.001 |
| MCV, FL         | 88.9 ± 13.5   | 81.4 ± 5.7    | 0.003   |
| MCH, pg         | 34.5 ± 3.6    | 31.0 ± 3.0    | < 0.001 |
| White cell count, \( \times 10^9 \) L | 11.4 ± 4.2 | 5.9 ± 2.1 | < 0.001 |
| Neutrophils, %  | 51.6 ± 11.3   | 47.9 ± 9.9    | NS      |
| Lymphocytes, %  | 38.8 ± 9.1    | 41.0 ± 9.5    | NS      |
| Platelets, \( \times 10^9 \) L | 409.1 ± 121.5 | 328.5 ± 95.6 | 0.002   |
| Systolic BP     | 109.5 ± 10.4  | 108.5 ± 9.6   | NS      |
| Diastolic BP    | 67.4 ± 8.4    | 70.4 ± 8.3    | NS      |

#### Blood transfusion history, n (%)

|                | Treated (n = 25) | Untreated (n = 14) | p-value |
|----------------|------------------|--------------------|---------|
| Yes            | 32 (82.1)        | 1 (2.8)            | < 0.001 |
| No             | 7 (17.9)         | 33 (97.1)          | NS      |

#### Folic acid supplement

|                | Treated (n = 25) | Untreated (n = 14) | p-value |
|----------------|------------------|--------------------|---------|
| Yes            | 38 (97.4)        | 1 (2.8)            | < 0.001 |
| No             | 7 (17.9)         | 33 (97.1)          | NS      |

#### Hydroxyurea use

|                | Treated (n = 25) | Untreated (n = 14) | p-value |
|----------------|------------------|--------------------|---------|
| Yes            | 25 (64.1)        | 0 (0.0)            | < 0.001 |
| No             | 14 (35.9)        | 0 (0.0)            | NS      |

#### Omega 3 fatty acids

|                | Treated (n = 25) | Untreated (n = 14) | p-value |
|----------------|------------------|--------------------|---------|
| Yes            | 4 (10.3)         | 0 (0.0)            | NS      |
| No             | 35 (97.3)        | 0 (0.0)            | NS      |

### Table 4: Anthropometric and hematological characteristics of the hydroxyurea treated and untreated (HbSS) groups.

| Characteristics | Treated (n = 25) | Untreated (n = 14) | p-value |
|----------------|------------------|--------------------|---------|
| Age, years     | 18.9 ± 2.4       | 19.3 ± 3.1         | NS      |
| Weight, kg     | 39.7 ± 9.7       | 37.7 ± 7.4         | NS      |
| Height, cm     | 151.4 ± 13.2     | 150.3 ± 8.5        | NS      |
| Body mass index, kg/m² | 17.50 ± 5.0 | 16.6 ± 2.5 | NS |
| Systolic BP    | 109.0 ± 10.4     | 110.4 ± 10.5       | NS      |
| Diastolic BP   | 65.4 ± 8.2       | 71.1 ± 7.9         | NS      |
| Hb, g/dL       | 9.2 ± 1.8        | 9.2 ± 1.1          | NS      |
| PCV, %         | 25.0 ± 7.3       | 25.2 ± 3.8         | NS      |
| MCV, FL        | 92.0 ± 6.3       | 83.4 ± 20.2        | NS      |
| MCH, pg        | 35.2 ± 3.5       | 33.2 ± 5.6         | NS      |
| White cell count, \( \times 10^9 \) L | 412.2 ± 130.2 | 403.0 ± 108.8 | NS |

#### History of blood transfusion, n (%)

|                | Treated (n = 25) | Untreated (n = 14) | p-value |
|----------------|------------------|--------------------|---------|
| Yes            | 20 (80.0)        | 12 (85.7)          | NS      |
| No             | 5 (20.0)         | 2 (14.2)           | NS      |

#### Folic acid supplement, n (%)

|                | Treated (n = 25) | Untreated (n = 14) | p-value |
|----------------|------------------|--------------------|---------|
| Yes            | 25 (100)         | 13 (92.8)          | NS      |
| No             | 0 (0.0)          | 1 (7.2)            | NS      |

NS: not significant; Hb: hemoglobin; PCV: packed cell volume; MCV: mean corpuscular volume; MCH: mean corpuscular hemoglobin; BP: blood pressure. Data presented as mean±SD unless otherwise indicated.
control women. These findings are consistent with the results of previous studies. In contrast to our findings, high BMI and obesity in American children and adults with SCA have been reported. It appears the anthropometric deficits commonly observed in patients with the disease in developing countries can be rectified by appropriate nutritional care and clinical management, particularly during childhood and adolescence.

The sickle cell patients who participated in this study were at steady-state, and some claimed to be on hydroxyurea (64.1%), folic acid (97.4%), and omega-3 fatty acids (10.3%). However, their hematological profiles, which were abnormal, were significantly different from that of the healthy control women. This finding was unexpected since hydroxyurea is known to improve hematological abnormalities in SCD. It is plausible the patients might not have been taking hydroxyurea as prescribed by their physicians because of its side effects (constipation/diarrhea, hair loss, muscle and joint pain, etc.), financial difficulties (patient have to pay for their medication in Sudan), inability to understand prescription guidelines and/or the drug became unavailable in private and hospital pharmacies. Indeed, the hematological data of the hydroxyurea treated and untreated patients, which were similar, reveals that the patients were unlikely to have been taking the medication as per guidance. Since most sickle cell patients in the country are poor. The government should explore the possibility of partially or wholly subsidizing hydroxyurea. Also, doctors must regularly check that their patients are following the prescription guidelines.

Small sample size (more patients who fulfill the inclusion and exclusion criteria could not be found in the same clinic), failure to collect quantitative nutrients intake data (most of the subjects were illiterate and they were unable to record their dietary intakes reliably), and failure to perform hemoglobin F tests (lack of laboratory facility) were the main limitations of the study. Regardless of these limitations, the study has provided a good picture of the nutritional and health status of Sudanese women of childbearing age with SCA.

CONCLUSION

The low anthropometric measures (height, weight, and BMI) and abnormal hematological values of the women with SCA in steady-state reflect sustained nutrition insults inflicted by the disease and poverty. Tailored nutritional counseling/advice must be an integral part of managing patients with SCA. Such advice is particularly vital for women of childbearing age because of the adverse effects of prepregnancy nutritional deficiency on birth outcomes.

Disclosure

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