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# Nutritional Profile of Children with Major Sickle Cell Syndrome at the Centre of Medical and Health Advice of Kipé, Conakry, 2018

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## Abstract

**Introduction:** Growth is a reflection of a child's health and nutritional status. Children with sickle cell disease often have slower statural and weight development. The aim of this study was to evaluate the nutritional profile of children with sickle cell disease (SCD) registered in the CEMECO centre database. **Methodology:** This was a cross-sectional study with simple random sampling of children aged 1 to 16 years registered in the clinic database. **Results:** We collected information on 208 children, 121 of whom had sickle cell disease and 87 of whom were normal, with a sex ratio of 1.02. The mean age of the sickle cell patients was 8.7 ± 4.4 years, while that of the non-sickle cell patients was 9.5 ± 4 years. Haemoglobin electrophoresis revealed 103 homozygous (SS), 18 double heterozygous (SC, SBetaThal, SE) and 87 normal (AA) and/or sickle cell trait (AS) sickle cell cases. We observed a significant difference in the height/age ratio ($P < 0.001$) and weight/age ratio characterising underweight ($P = 0.014$), expressed as a Z score, between the sickle cell patients and the non-sickle cell patients. On the other hand, for children over 5 years of age, the BMI and average income of sickle cell disease sufferers was similar to that of non-sickle cell disease sufferers ($P = 0.123$¥). **Conclusion:** The results of our study revealed stunted growth in children with sickle cell disease.

## Keywords

Sickle Cell Disease, Nutrition, Growth, Puberty, Guinea

## 1. Introduction

Growth is a reflection of a child's health and nutritional status. It is a biological process corresponding to an increase in the dimensions, weight, height, cranial and brachial perimeters of human beings [1]. Risk factors such as disease are not taken into account in normative representations of growth curves [2].

Sickle cell anaemia is the most common haemoglobinopathy, characterised by chronic haemolytic anaemia, with the homozygous form (SS) leading to severe, often fatal anaemia. This hereditary disease is responsible for complications such as vaso-occlusive crises, chronic haemolytic anaemia and recurrent infections, all of which affect children's growth and weight [3]. Reduced growth and delayed development are common in children with SCD, to the extent that some researchers advocate the use of modified growth curves to monitor their nutritional status. In general, these patients experience a progressive decrease in growth rate until adolescence. Micronutrients, such as serum zinc, 25-hydroxyvitamin D (25-OHD), selenium and retinol, are sub-optimal in people with SCID due to reduced appetite and inadequate food intake, increased energy and metabolic requirements and reduced physical activity [4].

These chronic manifestations combine delayed height and weight, and nutritional deficiencies in folates (which are renewed very rapidly during anaemic crises, thus depleting the folate stock) [5]. The prevalence of underweight in American children with sickle cell disease is 41% in cases of moderate malnutrition and 25% in cases of severe malnutrition, with a prevalence of wasting of 11% [6].

In the Congo, according to A. B. M'Pemba *et al*., 38% of female patients aged between 14 and 18 showed no signs of puberty and, at 16, 37% were still in their teens [7].

In Guinea, sickle cell disease is a major public health concern, and its prevalence is high, with an estimated 20% of the population carrying the sickle cell trait (EDS 2018) [8]. Several studies carried out in the paediatrics department of the Donka national hospital have addressed different aspects of this hereditary disease, but none has focused on the nutritional aspect of sickle cell disease. We felt it appropriate to study this report, the aim of which is to assess the nutritional profile of homozygous sickle-cell anaemia (SS) children in the database of the Laboratory Centre Medical et Conseils en Santé (CEMECO).

## 2. Methodology

### 2.1. Study Setting

This study was conducted at the Centre Medical and Conseils en Santé (CMCS) in Guinea. The choice of this centre was justified by the fact that it had a database of all sickle cell and non-sickle cell patients for whom haemoglobin electrophoresis had been performed.

Type and duration of study: we carried out a cross-sectional and analytical study of children and adolescents aged 1 - 16 years registered in the database of

the Medical and Health Advice Centre. The data were collected between 10 August and 25 December 2018.

From this database, we identified two groups of children, whose parents were called and scheduled at the center. Information on the sociodemographic and clinical characteristics of these patients was obtained using a questionnaire.

Group I (Exposed): all children aged 1 to 16 years registered in the centre's database with homozygous sickle cell disease (SS) confirmed by haemoglobin electrophoresis.

Group II (Unexposed): children aged 1 to 16 years, not with homozygous sickle cell disease (SS) registered in the centre's database matched for age and sex.

We did not include in our study children and adolescents with or without sickle cell disease whose parents did not wish to respond to calls and/or whose parents objected to the study.

Using the SCHWARTZ formula, the sample size was calculated as follows:

Number of cases: $N = Z\alpha^2 x \frac{pxq}{i^2}$

$N$ = required sample size

$Z\alpha^2$ = accepted risk of error ( $Z\alpha^2$ = 1.96²) or 95% confidence level

$P$ = prevalence of the factor studied ($p$ = 13.5%)

$q = 1 - p$

$i$ = margin of error at 5% (standard value of 0.05) or precision ($i$ = 0.05 or 0.1 etc.)

$n$ = 179

$$n = (1.96)^2 \times \frac{0.135 \times (1 - 0.135)}{(0.05)^2} = 179.4436 \cong 179$$

Our sample size, with a 95% confidence interval, a precision of 5% and a prior proportion of 13.5%, would be 179 cases according to the SCHWARTZ formula.

This sample size plus 10% non-response was rounded to 200 children.

Anthropometric measurements

- Weight measurements: Weights were measured using SECA scales. For children under 2 years of age or who could not stand, we used a baby scale.
- Height measurement: height measurements were taken using graduated towers and a MUAC-type stadiometer: children under 2 years of age were measured lying down, while older children were measured standing up.
- Brachial and cranial perimeters: were measured using a tape measure.

## 2.2. Assessment of Children's Nutritional Status

- In children under 5 years of age:

The nutritional status of children under five will be assessed using three anthropometric indices calculated based on age, height and weight:

- **Height for age:** This reflects the growth achieved in recumbent or standing height for the child's age at a given visit. This indicator is used to identify stunting, which is defined as a z-score < −2 standard deviations below the median height-for-age of the reference population. Severe stunting is considered when this z-score is < −3 standard deviations below the height-for-age median of the reference population.
- **Weight for age:** This reflects the body weight in relation to the child's age at a specific date. This indicator is used to determine whether a child is underweight or severely underweight. When the z-score is < −2 standard deviations below the weight-for-age median of the reference population, the child is said to be underweight or low weight. Severe underweight is considered to be when the z-score is < −3 standard deviations below the weight-for-age median of the reference population.
- **Weight for height:** This is a growth indicator that links weight and height in the supine or standing position, and indicates wasting (z score < −2 standard deviations below the weight-for-height median of the reference population). It is also a useful indicator for detecting overweight (z-score > 2 standard deviations below the weight-for-height median of the reference population) and obesity (z-score > 3 standard deviations below the weight-for-height median of the reference population).

### 2.3. In Children Aged Over 5

For these children, three anthropometric indicators will be used to assess their nutritional status:

- **Height for age:** This reflects the growth achieved in recumbent or standing height for the child's age at a given visit. This indicator is used to identify stunting, which is defined as a z-score < −2 standard deviations below the median height-for-age of the reference population. Severe stunting is considered when this z-score is < −3 standard deviations below the height-for-age median of the reference population.
- **Weight for age:** This reflects the body weight in relation to the child's age at a specific date. This indicator is used to determine whether a child is underweight or severely underweight. When the z-score is < −2 standard deviations below the weight-for-age median of the reference population, the child is said to be underweight or low weight. Severe underweight is considered to be when the z-score is < −3 standard deviations below the weight-for-age median of the reference population.
- **BMI for age:** This is a reliable indicator of body fat. It is used in children over the age of five to indicate wasting. Overweight was defined as a z-score between +2 and +3 standard deviations. Obesity was defined as a z-score > +3 standard deviations. BMI was calculated by dividing weight in kilograms (kg) by the square of height in metres (m).

Socio-demographic data on children (age, sex, origin) and parents (age, marital status, level of education, occupation, sibling size), clinical characteristics of

children on admission: transfusion history, vaccination status, clinical signs on inclusion (pallor, jaundice, fever, splenomegaly, hepatomegaly, bone pain, joint pain, chest pain, abdominal pain, respiratory distress).

### 2.4. Analysis Plan

The data collected was exported to R statistical software for cleaning and recoding. A descriptive analysis of child and parent characteristics was carried out separately for each group. Height-for-age, weight-for-age, weight-for-height and BMI-for-age z-scores were calculated for each child. The z-score values for which a flag was identified were excluded according to WHO standards. Exclusion was done by z-score and not by child. The z-scores considered as flags were height for age < −6 or > 6, weight for height < −5 or > 5 and weight for age < −6 or > 5. The valid z-scores were then categorised as a binary variable.

Pearson's chi-square test or Fisher's exact test was used to compare categorical variables. Quantitative variables were compared using the Student's t-test or the Wilcoxon test, depending on the normality of the distribution.

Anthro and Anthro Plus software (V1.0.4, 2007) were used to plot and compare the curves with the WHO reference population.

Univariate and multivariate logistic regressions were performed to identify factors associated with malnutrition. Variables with a p-value ≤ 20% in the univariate analysis or considered clinically important were included in the multivariate analysis using the stepwise ascending method. The effects of the factors studied were quantified by the odds ratio (OR) and their 95% confidence interval (CI).

## 3. Results

In the course of our study, we called, interviewed and collected information from 208 children with or without sickle cell disease out of 6497, *i.e.* a frequency of 0.03.

The study population included a total of 103 homozygous sickle cell patients (SS), 18 composite heterozygous sickle cell patients (SC, S$\beta$-thalassaemia and SE) (Group I) and 87 normal (AA) and/or sickle cell trait (AS) patients (Group II); Their ages ranged from 1 to 16 years, with an average of 8.7 ± 4.4 years for group I and 9.5 ± 4.2 years for group II. Males predominated, with 105 cases (50.5%) compared with 103 females (49.5%), with a sex ratio of 1.02.

Most of the children studied (93.3%) were Muslims, 6.3% Christians and 0.5 others. 76.9% were from the Conakry region; 60% of the children were of the Peulh ethnic group.

49% (102) of the children were cared for by people who had only studied the Koran, and 29.8% (62) had attended university.

Familial sickle cell disease was present in 72.6% of the children examined; parental consanguinity was noted in 40.4% of cases, with a predominance of 1st degree in 69%.

Socioeconomic status: 33.2% (69) of patients examined were poor.

The average anthropometric parameters were respectively 26.3 ± 12.7 kg with

extremes of 6.3 - 67.4 kg for weight; an average height of 126.7 ± 24.9 cm with extremes of 68 - 174.5 cm; a mean brachial circumference of 17.7 ± 3.3 cm with extremes of 12 - 29 cm; a cranial circumference of 52.5 ± 2.6 cm with extremes of 37 - 60cm and a body mass index of 15.5 ± 2.7 kg/m$^2$ with extremes of 11 - 36 kg/m$^2$.

Of the 48 children under 5 years of age, we found a mean Z-score P/T ratio of −0.64 ± 1.3 with extremes of -3.01 - 2.86, and a Z-score Weight/Age ratio of −0.63 ± 1.12 (extremes: −3.17 - 1.75), as well as a PB/PC ratio in Z-score at 0.3 ± 0.03 and extremes of 0.24 - 0.37; the mean Height/Age ratio in Z-score was performed on the entire study population (208 cases) and we found −0.74 ± 1.58 with extremes of −7.29 - 4.05. The BMI/age ratio was assessed in children aged 5 years and over; thus, out of 160 children assessed we found a mean of −0.84 ± 1.59 and extremes of −7.29 - 3.32. Of the total study population (208 children), 72.6% (151) had at least one family member with sickle cell disease; 40.4% (84) had parents who were related by blood; 32.2% (67) had been transfused, with an average number of transfusions of 2.73 ± 3.49 and a hospitalisation rate of 29.3% (61); 20.7% (43) had had jaundice at least once in their lives and 33.2% (69) had developed hand-foot syndrome.

During the study period 42.8% (89) of the children examined were pale, 23.6% (49) icteric, 96.2% (200) had adenopathy, 16.8% (35) hepatomegaly, 15% (31) splenomegaly and 19.7% (41) osteoarticular pain. The mean temperature of the population examined was 37.1˚C ± 0.59˚C with extremes ranging from 36.1˚C to 38.9˚C.

In this series of studies, the diagnosis of vaso-occlusive crisis was made in 21 children examined (10.1%), and osteoarticular pain syndrome in 17 children (8.2%). 1.4% of children developed clinical anaemia. Of the one hundred and three (103) children who underwent check-ups during the various consultations, we found an average haemoglobin level of 9.29 ± 2.9 g/dl with extremes of 2.9 to 14.6 g/dl; the average white blood cell count was 10.51 ± 5.79 G/l with extremes of 2.7 and 36 G/l; the mean red blood cell volume was 77.25 ± 11.46 fL with a range of 31.5 - 99.8 fL; the mean C-reactive protein count was 28.37 ± 29.94 mg/l with a range of 0.7 - 99.7 mg/l and the mean platelet count was 378.31 ± 196.35 G/l with a range of 30.9 - 999.9 G/l.

In the present study, we observed that the average weight of children in group I (24.0 ± 11.8 kg with extremes of 6.3 and 67.4 kg) was lower than those in group II (29.5 ± 13.2 kg with extremes of 9.2 and 64.1 kg); and this difference was statistically significant (P = 0.002¥).

Children in group I had a mean height (122.3 ± 24.5 cm with extremes of 68.0 and 174.5 cm) strictly less than those in group II (132.9 ± 24.2 cm with extremes of 82.5 - 174.4 cm) and this difference was statistically significant (P = 0.002¥).

The BP of children in group I was smaller than those in group II, and this difference was statistically significant (P = 0.001¥); on the other hand, the difference in body mass index (BMI) for children over 5 years of age was not statistically significant (P = 0.188¥), **Table 1**.

Table 1. Average staturo-weight growth in relation to the profile of respondents.

| Anthropometric parameters | Number of people with sickle cell disease (n = 121) "Group I" | Non-sickle cell population (n= 87) "Group II" | P-Value |
|---|---|---|---|
| **Weight, average (kg)** | 24.0 ± 11.8 (6.3 - 67.4) | 29.5 ± 13.2 (9.2 - 64.1) | 0.002¥ |
| **Height, average (cm)** | 122.3 ± 24.5 (68.0 - 174.5) | 132.9 ± 24.2 (82.5 - 174.4) | 0.002¥ |
| **PB, average (cm)** | 16.9 ± 3.1 (12.0 - 28.5) | 18.9 ± 3.3 (13.6 - 29.0) | 0.001¥ |
| **CP, average (cm)** | 52.1 ± 2.8 (37.0 - 60.0) | 53.2 ± 2.2 (47.0 - 58.2) | 0.085¥ |
| **BMI, average (kg/m²)** | 14.7 ± 2.9 (11.4 - 35.7) | 15.9 ± 2.6 (11.5 - 26.0) | 0.188¥ |
| **Weight/height, average (Z-score)** | −0.82 ± 1.1 (-3.01 - 1.62) | −0.29 ± 1.4 (-2.91 - 2.86) | 0.155¥ |
| **Weight/Age, average (Z-score)** | −0.91 ± 1.1 (-3.17 - 1.75) | −0.12 ± 1.0 (-2.43 - 1.59) | 0.014¥ |
| **Height/Age, average (Z-score)** | −1.1 ± 1.6 (-7.3 - 3.32) | −0.2 ± 1.3 (-5.2 - 4.05) | 0.000¥ |
| **BMI/Age, average (Z-score)** | −1.43 ± 1.16 (-3.9 - 1.57) | −0.99 ± 1.27 (-3.9 - 2.69) | 0.022¥ |
| **PB/PC average** | 0.29 ± 0.02 (0.24 - 0.34) | 0.32 ± 0.02 (0.27 - 0.37) | 0.009¥ |

¥ = Means test; Φ = Chi-square test.

We also observed that in children under five years of age, the mean weight/height ratio for children in group I was −0.82 ± 1.1 Z-score with extremes of −3.01 and 1.62 Z-score, which was lower than that for children in group II −0.29 ± 1.4 Z-score with extremes of −2.91 and 2.86 Z-score, although this difference was statistically insignificant (P = 0.155¥). On the other hand, the mean height/age ratio was evaluated for the entire study population and we found respectively −1.1 ± 1.6 Z-score with extremes of −7.3 and 3.32 in the exposed population and −0.2 ± 1.3 Z-score as well as extremes of −5.2 and 4.05 Z-score in the unexposed population, resulting in a statistically significant difference (P < 0.001¥). In children over five years of age, the mean BMI-for-age of exposed children was significantly lower than that of unexposed children, as shown in Table 1, and this difference was statistically significant (P = 0.022¥). The mean weight insufficiency (weight/age) of the exposed subjects was lower than that of the unexposed subjects, with respectively −0.91 ± 1.1 Z-score (extremes −3.17 and 1.75) and −0.12 ± 1.0 Z-score (extremes −2.43 - 1.59); this difference is statistically significant (P = 0.014¥).

Compared with unexposed children, sickle-cell anaemic children had a much greater history of familial sickle-cell anaemia, with a percentage of 51.4% in exposed children and 21.2% in unexposed children that was statistically significant (P < 0.001ф); on the other hand, the notion of consanguinity was not significant (P = 0.418ф).

Although the unexposed patients had a history of transfusion, jaundice, hospitalisation and hand-foot syndrome, there was a statistically significant difference between them and the exposed patients (P < 0.001ф) (see Table 2).

**Table 2.** Background of respondents in relation to their profiles.

| Background | Number of people with sickle cell disease | Number of non-sickle cell population | P-Value |
|---|---|---|---|
| **Average income** | | | |
| **Family history of sickle cell disease** | 107 (51.4) | 44(21.2) | 0.000ϕ |
| **Consanguinity, n (%)** | 53 (25.5) | 31 (14.9) | 0.418ϕ |
| **Transfusion, n (%)** | 59 (28.4) | 8 (3.8) | 0.000ϕ |
| **Average number of bags** | 2.92 ± 3.7 (1 - 27) | 1.14 ± 0.38 (1 - 2) | 0.000¥ |
| **Jaundice, n (%)** | 39 (18.8) | 4 (1.9) | 0.000ϕ |
| **Hospitalisation, average** | 2.98 ± 2.24 (1 - 11) | 2.18 ± 1.66 (1 - 6) | 0.000¥ |
| **Hand-foot syndrome, n (%)** | 59 (28.4) | 10 (4.8) | 0.000ϕ |

¥ = Means test; ϕ = Chi-square test.

The frequency of consultations due to pallor is more marked in sickle cell major syndrome (exposed) than in normal/AS (unexposed) and this difference remains statistically significant (P < 0.001ϕ). Splenomegaly was observed only in sickle cell major (exposed) patients (P < 0.001ϕ). Hepatomegaly was significantly more observed in major syndrome patients 14.9% (31) than in normal/AS 1.9% (4) with a P < 0.001ϕ.

We also observed a higher mean frequency of osteoarticular pain in exposed patients 15.9% (n = 33) than in unexposed patients 3.8% (n = 8) and this difference was statistically significant with a P = 0.005ϕ.

The diagnoses of vaso-occlusive crises and anaemia were observed only in exposed patients (major sickle cell syndrome); the number of patients found was twenty-one (n = 21) (10.1%) and three (n = 3) (1.4%) respectively. However, the diagnosis of osteoarticular pain syndrome was made in four patients (1.9%) without sickle cell disease and 13 patients with major sickle cell disease. This difference remained statistically significant (P < 0.001ϕ).

The mean Hb level in the two groups was respectively 8.3 ± 2.1 g/dl with extremes of 2.9 and 14g/dl in exposed patients and 11.8 ± 2.02 g/dl with extremes of 5.9 - 14.6 g/dl; the difference is statistically significant (P < 0.001¥). The mean red blood cell volume was slightly higher in the unexposed population than in the exposed, at 79.0 ± 10.6 fL (range: 53.0 - 99.8 fL) and 76.5 ± 11.8 fL (range: 31.5 - 99.8 fL) respectively. However, this difference was not statistically significant (P = 0.333¥).

The mean leucocyte count was more marked in the "exposed" sickle cell patients (11.9 ± 6.1 G/l with extremes of 2.7 - 36.0 G/l) than in the "unexposed" non-sickle cell patients (7.05 ± 2.8 G/l, extremes of 3.4 and 15.5 G/l) and this difference remained statistically significant (P < 0.001¥).

There was a slight difference in mean platelet counts between exposed and

unexposed subjects, with values of 394.5 ± 208.6 G/l (range 30.9 - 999.9 G/l) and 338.6 ± 158.9 G/l (range 77.1 - 728.0 G/l) respectively. However, this difference was not significant (P = 0.198¥).

Similarly, the mean level of C-reactive protein in the two study groups also showed a statistically non-significant difference (P = 0.101¥). The respective values were 33.1 ± 32.1 mg/l with extremes of 5.0 and 99.7 mg/l in major sickle cell patients and 15.0 ± 16.2 mg/l with extremes of 0.7 and 48 mg/l in normal patients (Table 3).

**Table 3.** Distribution of children according to clinical signs, diagnosis and paraclinical assessments.

| Variables | Effectiveness of major sickle cell syndrome (presentation) | Effective normal syndrome/AS (not exposed) | P-value |
|---|---|---|---|
| **Signs Clinics** | | | |
| Pallor | 82 (39.4) | 7 (3.4) | <0.01Φ |
| Jaundice | 46 (22.1) | 3 (1.4) | <0.01Φ |
| Splenomegaly | 31 (14.9) | 0 (0.0) | <0.01Φ |
| Hepatomegaly | 31 (14.9) | 4 (1.9) | <0.01Φ |
| Lymphadenopathy | 117 (56.3) | 83 (39.9) | 0.05f |
| Earache | 10 (4.8) | 13 (6.2) | 0.26Φ |
| Pain osteoarticular | 33 (15.9) | 8 (3.8) | <0.01Φ |
| Pain abdominal | 18 (8.7) | 8 (3.8) | 0.46Φ |
| Pain thoracic | 17 (8.1) | 3 (1.4) | 0.03Φ |
| Distress respiratory | 10 (4.8) | 4 (1.9) | 0.47Φ |
| Presence of rattles | 3 (1.4) | 3 (1.4) | 0.52Φ |
| Breath | 5 (2.4) | 2 (1.0) | 0.22Φ |
| Deficit engine | 0 (0.0) | 3 (1.4) | 0.12Φ |
| Achievement articular | 13 (6.2) | 4 (1.9) | 0.28Φ |
| Fever | 10 (4.8) | 34 (16.4) | <0.01Φ |
| **Diagnostic** | | | |
| Asthma | 0 (0.0) | 2 (1.0) | |
| Vaso-occlusive crises | 21 (10.1) | 0 (0.0) | <0.01Φ |
| Anemia | 3 (1.4) | 0 (0.0) | <0.01Φ |
| DOA Syndrome | 13 (6.2) | 4 (1.9) | <0.01Φ |
| Pneumonia | 1 (0.4) | 1 (0.4) | - |
| Pleurisy | 1 (0.4) | 0 (0.0) | - |
| Malaria | 3 (1.4) | 17 (8.2) | - |

**Continued**

| **Balance sheets paraclinics** | | | |
|---|---|---|---|
| THB | 8.3 ± 2.1 | 11.8 ± 2.02 | 0.000¥ |
| Hematocrit | 25.3 ± 6.8 | 33.7 ± 8.3 | 0.000¥ |
| white blood cells | 11.9 ± 6.1 | 7.05 ± 2.8 | 0.000¥ |
| Platelets | 394.5 ± 208.6 | 338.6 ± 158.9 | 0.198¥ |
| VGM | 76.5 ± 11.8 | 79.0 ± 10.6 | 0.333¥ |
| TCMH | 26.4 ± 6.5 | 26.7 ± 3.2 | 0.808¥ |
| CCMH | 38.8 ± 44.1 | 33.7 ± 3.6 | ¥0.540 |
| CRP | 33.1 ± 32.1 | 15.0 ± 16.2 | 0.101¥ |
| VS at the 1st Hour | 30.9 ± 22.6 | 19.9 ± 12.8 | 0.216¥ |
| VS at the 2nd Hour | 39.6 ± 24.7 | 39.1 ± 20.8 | 0.966¥ |

## 4. Discussion

Many of the children under 5 were stunted, acutely malnourished and underweight.

Comparison of anthropometric parameters (weight, height) between exposed and unexposed children showed a clearly significant difference, with P values of 0.002¥ and 0.001¥ respectively (see Table 1). A.B. M'PEMBA *et al.* in Congo reported in their study a clearly significant difference in weight and height between sickle-cell patients and controls (P = 0.00021) and (P = 0.0001) [7].

However, it should be noted that our results for body mass index (BMI) were not statistically significant (P = 0.188¥). A.L Saqladi *et al*., in their study of the growth and nutritional status of children with homozygous sickle cell disease, reported that children with sickle cell disease who received a regular transfusion over 2 years had a significant improvement in height, weight and BMI, with a Z close to normal [9].

Our results showed that in under-fives the mean weight/height ratio −0.82 ± 1.1 Z-score of sickle cell children is slightly lower than that of non-sickle cell children (−0.29 ± 1.4 Z-score) although this difference remains statistically insignificant (0.155¥). Unlike M.Y.P. Shongo *et al.* in 2015 in their study who had reported a statistically significant difference with a P < 0.05 [10]. This difference can be justified by the fact that our sample was larger than theirs.

Table 1 shows a statistically significant difference in the height/age ratio for the entire study population with a P < 0.001¥ and a mean BMI/age for sickle cell patients of −1.43 ± 1.16 vs. −0.99 ± 1.27 for non-sickle cell patients; this difference remains statistically significant (P = 0.022¥). S.E. Cox *et al.* in Tanzania in 2011 reported height deficits in sickle cell patients during adolescence, as well as a prolonged growth period with P = 0.015 [11].

## 5. Strengths and Limitations of the Study

To our knowledge, this study is one of the first to analyse the relationship between the nutritional and pubertal profile of children with homozygous sickle cell disease (SS) and those without. It has the advantage of having made it possible to assess children in both groups throughout the study period. However, it may have limitations. It involved only 208 children, thus limiting the possibility of in-depth modelling of the data; the second limitation was linked to the absence of systematic blood tests for all participants.

## 6. Implications for Research and Practice

The results of this study contribute to a better understanding of the follow-up of children with sickle cell disease and the strategies used by doctors to adapt to the management of sickle cell disease. They will help guide actions to improve access to laboratory services. They will also make it possible to plan other, larger studies that will provide more important data for a better understanding of the disease.

## 7. Conclusion

At the end of this study, we found a statistically significant difference between the evolution of weight and height growth in children with major sickle cell syndrome and healthy children of the same age group. Chronic recurrent anaemia, vaso-occlusive crises, increased energy expenditure required for adaptation mechanisms and undernourishment are among the factors thought to explain this delay.

## Authors' Contributions

Study design: DMA, TA;
Data collection: DMA;
Data analysis: CSC, CAT, MAD;
Initial drafting of the manuscript: BMA, DM, DMA;
Manuscript revision: TA, BMA, DMA, DM, CSC, TAO.

## Availability of Data and Materials

The data supporting the results of this study are available from [DOUKOURE Mamadou Aliou], but restrictions apply to the availability of these data and are therefore not publicly accessible, as our team is working on other analyses using the same data, which will then be submitted for publication. However, these data are available on reasonable request from the corresponding author [DOUKOURE Mamadou Aliou].

## Consent for Publication

Not applicable.

## Conflicts of Interest

The authors declare that there are no competing interests.

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