Exploring the Nutritional Status of Stickler Syndrome Patients

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Abstract

Introduction: Growth retardation in weight and height was reported among patients with sickle cell anemia (SCA).The growth retardation, delayed sexual development and poor immunologic response are possibly due to the under nutritional state associated with the disease. The active metabolic state observed in sicklers is due to an increase in the synthesis of Hb, cardiac work, cell turnover, and a decrease in appetite and intake. Objectives: Tto compare anthropometric measurement (weight, height and body mass index (BMI)) between sicklers and normal children and to study factors related to the nutritional status of sicklers.

Patients and methods: This comparative hospital based study based study done among sicklers and control group using a questionnaire.

Results: Stunted weight (less than 3rd centile) was observed in 49 (56.3%) sicklers compared to 9(11.4%) controls, 38(43.7%) sicklers and 51(64.6%) controls had their weight in the range between 5th-95th centile while 30(34.5%) sicklers and 7(8.9%) controls had their height less than 5th centile. A total of 40(46%) sicklers and 53(67.1%) control had a height outside the range of than 5th-95th percentile ,however it was observed that 17(19.5%) sicklers had their height more than 95th compared to 19(24%) controls. BMI was recorded as less than 5thin 51(58.7%) of the sicklers compared to7(8.9%) in the controls. Children from low social class were recorded in79.6%, 63.3% and 78.4%of weight, height and BMI stunting respectively .Finally stunting in weight, height and BMI was more obvious in older children.

Conclusion: Most of sickle cell anemia patients had weight and body mass index retardation, this problem was mostly observed towards adolescent age.

Introduction

Sickle cell disease (SCD) is common in different parts of Sudan in general and in Misseria tribe in particular [(1)]. More than 5% of patients attending to Khartoum teaching clinks (Khartoum, Sudan) were sicklers, therefore SCD is the main hemoglobinopathy in Sudan [(2)]. Growth retardation among patients with chronic illness encourages many authors to suggest special growth cares for those patients. Many patients with chronic illnesses like syndromes and cystic fibrosis have well developed growth curve to match their problems [(3-5)]. Growth retardation in weight and height was reported among patients with sickle cell anemia (SCA) [(6)]. Sickle cell disease (SCD) is a type of hemoglobinopathy that leads to abnormal production of hemoglobin, called Hb SS [(7)]. The growth retardation, delayed sexual development and poor immunologic response are possibly due to the under nutritional state associated with the disease [(7)]. The active metabolic state observed in sicklers is due to an increase in the synthesis of Hb, cardiac work, cell turnover, and a decrease in appetite and intake. All of the above could be explained by the increase in needs and a demand for protein as well as calories [(8)]. Some studies among sicklers have established an increase in the catabolic process, a release of some inflammatory mediators and as a result an increase in the nutritional requirement. Growth retardation, particularly in weight and sexual maturation was observed by previous studies; however height might not be affected [(9, 10)]. The decrease in intestinal absorption, the increase in cell

turnover might result in a decrease in body mass and other anthropometric measurements[11, 12, 13].***Rationale:*** there is an urgent need to evaluate nutritional status of the patients suffering from SCA. Knowledge of the growth parameters of patients with SCA can stimulate pediatricians to design special growth curve for them ***Objectives:*** to compare anthropometric measurement (weight, height and body mass index (BMI)) between sicklers and normal children and to study factors related to the nutritional status of sicklers.

Patients and methods

This comparative hospital based study was conducted in Gineana general hospital, Gineana town, Western Sudan. Sickle cell anemia is known to be prevalent in that area. Gineana general hospital has a unit of pediatrics as well as other major medical specialty units and supportive services. The study was conducted in April 2011, over 3 weeks during a medical mission coordinated with the hospital by a group of doctors from different specialties. Ethical clearance was obtained from the research ethics committee of the hospital and patients consent to participate in the study was obtained from their parents. Inclusion criteria included the following: 1- age 7-15 years and known to be sicklers. 2-sickle cell anemia confirmed with hemoglobin genotype "SS" on hemoglobin electrophoresis. Every patient was assessed by the author. The control group was selected randomly from those children who attended the survey for other purposes. For both patients and controls, a focused history was obtained including personal, past medical, family and nutritional history. Two investigators independently measured the body weight to the nearest decimal of kilograms and the height to the nearest decimal in centimeters by using standard techniques (Two-in-One Beam Scale by Detecto[12]. The body mass index (BMI) was then calculated by dividing the bodyweight in kilograms by height in meters squared. Applying normal weighing scale and recording approximated to 0.1 kilogram, weight was carefully taken for sicklers and control in light clothes and in standing position unsupported by kilograms unit [12].Height was also estimated in CNM in standing position using normal stand meter with recording up to 0.1cm [12]. A diagnosis of stunted weight, height or BMI was based on WHO standard growth charts for body weight, length or height by conventional methods. The formulae used to calculate body mass index is by dividing weight in kilogram divided by height in meter according to WHO and Centers for Disease Control classification. Patients or controls are considered underweight when weight was less than the 5th percentile. Normal weight was considered when the BMI was from 5th up to the 85th percentile. Overweight is considered when the BMI is from 85th to less than the 95th percentile [15, 16]. A modified Kuppuswamy's Socioeconomic Scale was used to estimate the socioeconomic status [17]. All data was entered in Excel 2003 program and descriptive statistics was calculated. Chi Square test was used to compare dichotomous variables and tor Z test for test of proportions. A p value of 0.05 was used to determine significance.

Results

In this study, two groups were enrolled; 87 patients and 79 controls. Males in the patients and control groups were 49, 43, respectively; female in the patients and control groups were 38, 36, respectively .There was no significant difference between males and females in patients and control groups, where p- value = 0.807 (Table 1) .7 and 9 from the patients and the control groups were in the upper social class, respectively whereas10 and 9 from the patients and control groups were from the upper middle social class, respectively. From all 15, 17 the patients and the control groups were from Lower middle social class, respectively. Twenty-five and 20 from the patients and the control were from the lower social class. No significant differences were observed in cases and control groups among the different social classes (p>0.005) (Table 2). Thirty nine (79.6%), 19(63.3%) and 40(78.4%) of sicklers who suffered from weight, height and BMI stunting were from the low social class with (p= 0.107). eight (16.3%), 11(36.7%) and 11(21.6%) of sicklers who suffered from weight, height and BMI stunting were from the low social class (p=0.04) was observed for weight ,height and BMI for sicklers in lower middle class (Table 3). Regarding weight; 49%, 9% from the patients and the control groups had their weight below 5th centile respectively. Thirty eight and 51 from the patients and the controls had weight more than 5th-95thpercentile respectively. Nineteen of controls had a weight more 95th centile (Table 4). Regarding height; 30 and 7 of the patients and the controls had height below 5th centile, respectively while 40 and 53 from the patients and the control had height between 5th -95th centile, respectively. Seventeen and19 from the patients and the control had height above 95th centile respectively (Table 4). Body mass index (BMI) of 51 of the sicklers and 7 of the control was less than 5th centile respectively. Thirty-five of the sicklers and 67 of the control had BMI less than 5th- 85thcentilerespectively.Oneof the sicklers and 5 of the control had BMI more than 85th centile respectively (table 4). Stunted weight, height and BMI in sicklers was highly significant (p value = 0.005) (table 4).The deterioration in weight, height and BMI increase when children advanced in age, however this was not statistically significant (p > 0.005) (Table 4).

Stunted weight(less than 3rd centile) was observed in 49(56.3%) sicklers compared to 9(11.4%) control group. 38(43.7%) sicklers and 51(64.6%) control had their weight in the range between 5rd -95th. 30(34.5%) sicklers and 7(8.9%) control had their height less than 5d, 40(46%) sicklers and 53(67.1%) control their height between More than 5rd -95th, however it was observed that 17(19.5%) sicklers had their height more than More 95th compared to 19(24%) control group. BMI was recorded as Less than 5rdin 51(58.7%) of the sicklers compared to7(8.9%) in the control group. 79.6%, 63.3% and 78.4%who suffered from weight, height and BMI stunting were from low social class Finally stunting in weight, height and BMI more obvious in older children.

In the current study, it was observed that children with sickle cell anemia (SCA) were stunted in weight (56.3%), height (34.5%) and their body mass index (58.7%). These findings are in harmony with those of other studies. In Sudan , studies by Yasin Haj, (2004), Sara Hashim (2015) agree with our findings regarding weight but disagree regarding height[17,18].There were similar studies done in Nigeria by Esezoboretal (215), and in Tanzania by Cox et al.(2012) among sicklers within the same age[19,20]. Similarly, Al-Saqladi·s findings of stunting and wasting accounted to 54% and 35% are in support of our findings in children with SCA[21]. Our results were consistent with studies in Pakistan by Odetunde et al. (2016) together with Singhal et al[22,23]. In developed countries like USA, Chawla et al [24], Chawla et al [25], sicklers were reported to be obese as well as overweight. The social and health fairness in USA regarding health, social and psychological facilities can ultimately lead to obesity and overweight. In developing countries, shortage of resources, limitation of facilities, deficiency in drug and vaccinations, repeated infection and social issues lead to concerns regarding physical, sexual and developmental problems. In this study, height is less affected than weight, a finding which is consonant with other studies [26, 27].These findings might be related to glands function or age of sexual maturation. More stunting in weight, height and BMI was observed in older children between 12-15, which agreed with Kawchak et al [28]. Of course, chronicity of the problem will lead to clear complications with advancement in age. It was observed that in this study most of the stunted sicklers were among those in low middle social class. This was also supported by Wolf et al. in developing countries [29]. Health care accessibility, reasonable income and other facilities make sicklers from higher classes suffer less than those from low classes.

Limitations

The numbers of cases and control is small. Diet and nutritional habit were not considered

Conclusion

Most of sickle cell anemia patients had weight and body mass index retardation, this problem was mostly observed towards adolescent age.

Recommendations

More efforts regarding quality of care for sicklers regarding nutritional, social and psychological are highly and urgently needed. Multi centre study should be encouraged to study the quality of life among sicklers.

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Conflicts of interest

No conflict of interest

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Table 1: Gender distribution

Gender	Patients	Control	Total
Males	49	43	92(55.4)
Females	38	36	74(44.6)
Total	87	79	166(100)

Chi square value = 0.060 p- value = 0.807 (not significant differences among the gender for cases and control)

Table 2: Socioeconomic status (according Kuppuswamy scale) comparison between sicklers (n 87) and control (79) according Kuppuswamy scale

Social class	Patients	Control	Total	p-value
Upper	7	9	16(14.3%)	0.509
Upper middle	10	9	19(17%)	0.936
Lower middle	15	17	32(28.6%)	0.483
Lower	25	20	45(40.2%)	0.492
Total	87	79	166 (100)	

No significant difference between cases and control regarding social classes

Table 3: Socioeconomic status (according Kuppuswamy scale)among sicklers who are stunted in weight (n 49), height (n 30), BMI(n 51)

Social class	p- value	Stunted weight	Stunted height	Stunted BMI
Upper	N/A	0	0	0
Upper middle	0.267	2(4.1	0	0
Lower middle	0.04	8(16.3%)	11(36.7%)	11(21.6%)

Lower	0.107	39(79.6%)	19(63.3%)	40(78.4%)
Total		49(100)	30(100)	51(100)

It is significant for stunted growth, height and BMI to be associated with lower middle class

Table 4:Comparison in anthropometric measurement between sicklers (n 87) and control (79)

	Patients	**Control**	**Total**	**p-value**
Weight Centile				
Less than 5rd	49(56.3%)	9(11.4%)	58	0.0001
More than 5rd -95th	38(43.7%)	51(64.6%)	89	0.0006
More95th	0(0.0%)	19(24.0%)	19	0.0001
Total	87(100 %)	79(100 %)	166	
Height Centile				
Less than 5^{d}	30(34.5%)	7(8.9%)	37	0.001
More than 5rd -95th	40(46%)	53(67.1%)	93	0.04
More 95th	17(19.5%)	19(24%)	36	0.483
Total	87(100%)	79(100%)	166 (100)	
Body Mass Index (BMI) distribution of the subjects (HbSS) and controls (HbAA)				
Less than 5rd	51(58.7%)	7(8.9%)	58	0.001
More than 5rd - 85th)	35(40.1%)	67(84.8%)	102	0.001
More 85th	1(1.1%)	5(6.3%)	6	0.07
Total	87(100%)	79(100%)	166	
Relationship between anthropometric measurement and age among cases (Age/Measurement)	**7-11 year**	**More than11-15 year**		
Weight Less than 5rd	21	28	49	0.118
height Less than 5rd	14	16	30	0.48
BMI	20	31	51	**0.012**