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Original Article

Blood Pressure in Children with Sickle Cell Disease in a Tertiary Hospital, Sokoto, Nigeria

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A R T I C L E I N F O

Keywords:
Blood pressure,
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A B S T R A C T

Background: Sickle Cell Disease (SCD) is a genetic disorder and the most common form of haemoglobinopathies. It has 2 main pathological processes,vaso-occlussion and haemolysis. These lead to complications including haematuria, proteinuria, concentrating defect, renal insufficiency and hypertension with consequent progression to chronic kidney disease (CKD.Studies have reported that there were significantly lower blood pressures among children with SCD than those with normal heamoglobins.This study aimed to determine the systolic (SBP) and diastolic (DBP) blood pressures of sickle cell disease patients in steady state compared with normal HbAA controls in a tertiary hospital in Sokoto, Nigeria. Methodology:A cross-sectional study among diagnosed SCD patients in steady state, 6 months – 15 years, attending thepaediatrichaematology clinic of the UsmanuDanfodiyo University Teaching Hospital, Sokoto, and age matched controls with normal HbAA attending Peadiatric Outpatient Clinic. Clinical data obtained were socio-economic status of the parents, patient's age, gender, past medical history (numberof blood transfusion and crises/year) and routine medications. Weight, height/length and blood pressure measurement of the subjects were documentedwith their respective packed cell volume andhaemoglobinelectrophoretic pattern. Results: Fifty-five (100%) recruited SCD patients have normal SBP and DBP, out of the 50 (100%) controls, 5/50(10%) have systolic hypertension and 19/50 (38%) have diastolic hypertension. The mean SBP and DBP for SCD were 94.62 ±10.13SD and 58.43 ±10.49SD and that of the controls were 99.53 ±14.90SD and 69.25 ±12.42SD each respectively. The mean SBP between the subjects and controls is not significant, (p=0.616) as against the positive significant difference in the mean values of DPB between the two. P<0.05. Conclusion: The mean values for both SBP and DBP recorded in SCD patients are lower compared to that of the controls. More studies needed particularly on patients with sickle cell nephropathy to determine those with hypertension and CKD.

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Introduction

Sickle Cell Disease (SCD) is the most common forms of haemoglobin disease [1]. It is a genetic disorder associated with structurally abnormal haemoglobins resulting in the formation of sickle-shaped red blood cells and several clinical manifestations from two main pathological processes. These are vaso-occlussion and haemolysis [2,3,4].

The kidneys are highly vascularised organs and vulnerable to vaso-occlusive injury. The recurrent vaso-occlusion and haemolysis lead to functional and structural changes presenting as haematuria, proteinuria, concentrating defect, renal insufficiency and hypertension which may progress to chronic kidney disease (CKD)[4,5,6,7].

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Studies done on blood pressures in sickle cell anaemia patients reported that there were significantly lower blood pressures than their controls [1,8,9,10,11,12]. The exact reasons for lower systemic blood pressure in sicklers were not known, but certain pathophysiologic factors have been attributed, these includes; a medullary defect, hyperdynamic circulation due to anaemia, enhanced production of prostaglandins as well as nitric oxide and lastly, decreased vascular reactivity secondary to chronic vasculopathy, these causes lower systolic blood pressure and systemic vasodilatation respectively [1,8-12]. This study aimed at to determine the systolic and diastolic blood pressures of sickle cell disease patients in steady state compared with normal HbAA control in a teaching hospital Sokoto, Nigeria.

Materials and Method

A cross-sectional descriptive study carried out on diagnosed sickle cell diseasepatients aged 6 months – 15 years who were regularly attending thepaediatrichaematology clinic of the UsmanuDanfodiyo University Teaching Hospital, Sokoto, in their steady state and on routine drugs (folic acid, paludrine, hydroxyurea and penicillin V respectively) from October 2022 – March 2023. Steady state is defined as absence of any crisis in the preceding 4 weeks, and absence of any symptoms or signs attributable to acute illness.

All subjects diagnosed/confirmed HbSS and HbSC that met the inclusion criteria were recruited. Those subjects with congenital/acquired heart disease,renal disease, crises, recent blood transfusion, patients on antihypertensive and steroids were excluded. Controls were

apparently healthy, age matched patients with normal HbAA attending Peadiatric Outpatient Clinic for follow up after treatment for acute illness/minor ailments, fully recoveredand met the inclusion criteria.

Sample size of 50 subjects was used as determined and obtainable in previous studies.1,10,13Ethical approval was obtained from the Ethics and Research Committee of the hospital.

Questionnaires were administered to parents and subjects who consented and assented each respectively. Clinical data obtained were socio-demographics that include socio-economic status of the parents, patient's age, gender, past medical history (numberof blood transfusion/crises in a year) and routine medications. General physical examination, weight, height/length and blood pressure measurement of the subjects were documentedwith their respective packed cell volume (PCV) andhaemoglobinelectrophoretic pattern.

Anthropometry: Both subjects and the controls had their height/ length and weight measured using Mini meter portable device and stadiometer, respectively. Body mass index (BMI) wasobtained for all using the formula: BMI= Weight in kilogram / (Height in meter)2and then appropriately classified according to WHO growth charts.

Blood Pressure measurement: Astandard procedure was used in determining each patient BP according to the recommendation of the task force on high blood pressure in children and adolescent [14]. Measurements were made using a mercury sphygmomanometer with mining appropriate cuffstaken at two and or three different times, always by the same author, with minimal or no anxiety and fear in all patients. Hypertension was defined as average systolicblood pressure (SBP) and/ or diastolic blood pressure (DBP) equal or greater than 95th percentile for age, gender and height. Pre-hypertension in children was defined as average SBP or DBP ≥90th percentile, but less than 95thpercentile, while low BP was defined as a blood pressure value <5th percentile or below 2 standard deviation of themean for age and gender.

Data analysis: Statistical Packages for Social Sciences (SPSS Inc Chicago, IL, USA) software version 23.0 was used to analyzed the data obtained. The data were presented as means, standard deviation, frequencies and percentages. pvalue≤0.05 was taken as significant.

Results:

Fifty-five (52.4%) sickle cell disease (SCD) subjects in steady state were enrolled during the study period, their mean age was9.66±3.3SD, 26(47.3%) were males and 29 (52.7%) females with male: female ratio of 1:1.1. Out of the 55 subjects, 47 (85.5%) have HbSS pattern, 1 (1.8%) HbSS+F and 7 (12.7%)HbSC respectively.Fifty (47.6%) controlswere recruited with mean age of 6.54±4.59SD, 33(66%)males and 17(34%) females each respectively.The difference in the mean ages between the SCD patients and that of the controls is not statistically significant.(See Table I age specific and gender distribution).

Table I: Age specific and gender distribution

	Age SCD		Control		
	Male	Female	Male	Female	Total
<1	-		2	1	3
1-<5	2	3	11	5	21
5-<10	9	8	9	6	32
10-15	15	18	11	5	49
Total	26	29	33	17	105

The packed cell volume (PCV) of the SCD patients ranges from 16 - 35% with mean PCV of 26.65 ±4.27SD which was lower compared to that of the controls that ranges from 23 - 40 % and mean of 30.33±6.76SD. p=0.790. Paradoxically, the SCD patients has higher mean weight and height measurements, 25.67±10.51SD & 130.17±19.14SD but lower values were obtained for the controls,18.67±10.87SD & 109.35±24.38SD respectively. While both mean BMI values of the subjects and controls were low, subjects recorded 15.9 ±5.49SD and 15.1±1.98SD was recorded for the controls. The difference in weight, height and BMI of the Subjects and controls were not significant. P>0.05. (see Table II)

Table II: Mean clinical variables of the SCD patients and the Controls

Variable	SCD (n=55)	Control) (n=50)	p-value
PCV	26.65±4.27	30.33±6.76	0.184
Weight	25.67±10.51	18.67±10.87	0.093
Height	130.17±19.14	109.35±24.38	0.414
BMI	15.9±5.49	15.1±1.98	0.630
SBP	94.62±10.13	99.53±14.90	0.616
DBP	58.43±10.49	69.25±12.42	0.0001

Among the 55 subjects recruited in this study, 10 (18.2%) have normal weight, 45 (81.8%) were underweight and none was overweight nor obese. On the other hand, the controls have 11(22%) with normal weight, 37 (74%) were underweight and 2 (4%)overweight. (Table III)

Table III: BMI distribution in SCD and the Controls

BMI	SCD n (%)	Control n(%)
Normal	10 (18.2)	11 (22)
Underweight	45 (81.8)	37 (74)
Overweight	0	2(4)

All the 55 SCD patients have normal SBP and DBP, but for the controls, 5/50(10%) have systolic hypertension and 19/50 (38%) have diastolic hypertension. The mean values for both systolic blood pressure SBP and diastolic blood pressure DBP recorded in SCD patients are lower compared to that of the controls. For SCD, the mean SBP = 94.62 ±10.13SD, mean DBP = 58.43 ±10.49SD and the controls have mean SBP of 99.53 ±14.90SD and mean DPB = 69.25 ±12.42SD. There is no difference in the mean SBP between the subjects and controls (p=0.616) as against the positive significant difference in the mean values of DPBbetween the two. P<0.05.(Table II)

Twenty-four (43.6%) of the SCD patientshad previous history of blood transfusions over one year period (20 subjects had one transfusion, 2 had two transfusions and the remaining 2 had three or more transfusions). In addition, 40 (72.7%) of the subjects had previous history of Vaso-occlusive crisis (21 had an episode, 9 had 2 episodes while 10 had 3 or more episodes.).blood transfusion or frequencies of blood transfusion as well as crises does not significantly affect blood pressures among SCD patients. p>0.05.

Discussion:

Sickle cell disease is highly prevalent (11%) [15] in the study area. Increase in blood pressure among sickle cell disease may increase morbidity and mortality. Our study recorded lower mean SBP and DBP in SCD patients than the controls. Similar previous studies by Animashaun et al,[8] Aneke et al [9] and Faruk et at (Nigeria)[10], Pegelow et al (USA) [11], Homi et at (Jamaica)[12] reported the same pattern of lower mean values of SBP and DBP respectively. But other studiesGeorge et al,[13] Shatat et al,[16] Becker et al [17] &Ho Chi Hsien et al[18] recorded significantly higher mean SBP and DBP among SCA children than the controls. However, study by Ekure et al,[19] Smit et al and Desai et al [20] reported no significant difference in the mean blood pressures (SBP and DBP) between the SCD patients and controls.

Previous studies postulated the mechanisms that lower BP in SCDpatients to include renal tubular defects,hyposthenuria increased Sodium and water loss. Our study recorded low mean PCV and low BMI among the SCD, these can cause alterations in peripheral vascular

resistance and vasodilation with consequent affectation of cardiac output resulting low blood pressure.Again, our study did not record any significant difference in blood pressure among SCD subjects in relation to VOC/painful crises and blood transfusions. Similar findings were also obtained by Ammar et al,[21] DeBaun et al [22] and Lamarre et al [23] as no association was found between BP and a history of painfulVaso-occlusive crisis. But it has also been reported that the riskof occlusive stroke increases with SBP andno significant relationship between DBP and occlusive stroke was observed. No explanation as to why we observed negative relationship between painful crises/ blood transfusions and blood pressure among SCD patients, however it was documented that SCD children who are regularly transfused have significantly lower comorbidities.

Conclusion:

The mean values for both SBP and DBP recorded in SCD patients are lower compared to that of the controls. More studies needed particularly on patients with sickle cell nephropathy to determine those with hypertension and CKD.

CONFLICT OF INTEREST

The authors declare that no conflict of interest exists

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