



Hypertension in Children with Sickle Cell Disease: A Comparative Study from Port Harcourt, Nigeria

I. O. George^{1*}, P. N. Tabansi¹ and C. N. Onyearugha²

¹Department of Paediatrics, University of Port Harcourt Teaching Hospital, Nigeria.

²Department of Paediatrics, Abia State University Teaching Hospital, Nigeria.

Authors' contributions

This work was carried out in collaboration between all authors. Author IOG designed the study, wrote the protocol, and wrote the first draft of the manuscript. Authors PNT and CNO managed the literature searches, analyses of the study. All authors read and approved the final manuscript.

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ABSTRACT

Background: Sickle cell anaemia is a chronic anaemia that is characterized by episodes of severe bone pain from blood vessels occlusion by sickled red blood cells when deoxygenated, and eventual end organ affectation and multi-organ failure. The aim of this study was to compare the arterial blood pressures of children with sickle cell anaemia in steady state with those of age- and sex-matched healthy controls and to identify those with hypertension.

Materials and Methods: This cross-sectional descriptive study was conducted in the Outpatient Paediatric Haematology Clinic of University of Port Harcourt Teaching Hospital from January to March 2015. Blood pressure, weight and height were measured and a specific form was used to record data.

Results: There were a total of 50 children with sickle cell anaemia in stable state during the study period. Of these, 31 were male while 19 were females giving a Male: Female ratio of 1.6:1. All the patients had HbSS genotype. Most of them 22(44%) were between the ages of 5 and 10 years. The mean packed cell volume was 22.79±4.34. Majority of the patients had packed cell volume between 16 and 30. Most 41(82%) of them were underweight. The prevalence of hypertensive is 22%. Majority (82%) of them had low Body Mass Index.

Conclusion: There is no significant difference in the systolic blood pressure of children with sickle

*Corresponding author: E-mail: geonosdemed@yahoo.com;

cell anaemia compared to age and sex matched controls. Hypertension appears to be frequently undiagnosed by paediatric clinicians. Early, appropriate diagnosis is important so as to establish effective treatment for abnormal blood pressure.

Keywords: Blood pressure; children; sickle cell anaemia; body mass index; Nigeria.

1. INTRODUCTION

Sickle-cell disease is a genetic disorder of growing public health importance worldwide [1]. More than 300 000 homozygous neonates with sickle-cell anaemia are born every year, with three-quarters born in sub-Saharan Africa [2]. It is a chronic anemia that is characterized by episodes of severe bone pain from blood vessels occlusion by sickled red blood cells when deoxygenated, and eventual end organ affectation and multi-organ failure [1].

Hypertension in children may be secondary to another disease process or it may be essential hypertension. Secondary hypertension is more common in children than in adults, and common causes of hypertension in children include renal disease, coarctation of the aorta, and endocrine disease [3]. However, the majority of children and adolescents with mild to moderate hypertension have primary hypertension in which a cause is not identifiable. Hypertension in children has been shown to correlate with family history of hypertension, low birth weight, and excess weight [2,4,5]. Several studies [6,7] have shown that blood pressure is generally lower than normal in individuals with sickle cell anemia compared to age and sex matched controls. The exact mechanism of this lower blood pressure is unknown but factors such as salt losing sickle cell nephropathy have been implicated [7].

Hypertension is known to be more prevalent among people of the black race, who also frequently carry the sickle cell gene [2]. Sickle cell disease (SCD) is associated with high morbidity from recurrent episodes of vaso-occlusive and anaemic crises. Mortality most times occurs during acute crises and may be secondary to various organ failures including the kidneys. Kidney disorders are often associated with hypertension.

Comparative data on the blood pressure values of this subgroup with the normal population is scarce and mainly in the adult population of sicklers. Nevertheless, information on prevalence of hypertension among children with sickle cell anaemia in our environment is limited. This study

thus seeks to determine the blood pressure levels of children with sickle cell disease and compare them with those of normal reference values. This will identify children with hypertension and its resultant morbidity such as stroke.

2. MATERIALS AND METHODS

This cross-sectional descriptive study was conducted in the Outpatient Paediatric Haematology Clinic of University of Port Harcourt Teaching Hospital (UPTH) from January to March 2015. This study enrolled all patients with SCD (haemoglobinopathy HbSS and HbSC) aged 6months to 16 years, who were seen in the outpatient service during the study. Exclusion criteria included recent (within 2 weeks) hospitalization and/or episode of acute chest pain, pain crises, febrile illness or blood transfusion.

Blood pressure (BP), weight and height were measured and a specific form was used to record data. Three BP measurements were made for each patient at three different times, always by the same author, and all care was taken to minimize anxiety and fear of the procedures. Measurements were made using a mercury sphygmomanometer with appropriate cuffs. Systolic (SBP) and diastolic BP (DBP) were defined as normal when below the value of the 95th percentile for age, sex and height. Hypertension was defined as BP greater than the 95th percentile for age, sex and height percentile [8].

Weights would then be assessed using a Seca weighing scale, with the child completely undressed. Weights will be measured in kilograms, to the nearest 0.1 kg (100 grams). Seca weighing scale was used to measure weight, and a wooden vertical stadiometer was used for height; the horizontal rod was adjusted to rest on the top of the head at a right angle with the vertical ruler. Anthropometric measurements were made with the patient barefooted and wearing as little clothing as possible. Nutritional status according to Body Mass Index (BMI) was classified using the growth charts issued by the World Health Organization (WHO).

Ethical approval was obtained from the Ethical Committee of the UPTH.

Means, standard deviation, frequencies and percentages, correlation and analyses will be done in order to predict the significant values with p value of 0.05.

3. RESULTS

There were a total of 50 children with sickle cell disease in stable state during the study period. Of these 31 were male while 19 were females giving a Male: Female ratio of 1.6:1. All the patients had HbSS genotype. There were more children 22(44%) between 5-10years as shown on (Table 1). The mean packed cell volume was 22.48 ± 4.1 (range 15-32) (Table 2). The

frequency distribution of their packed cell volume (PCV) is as shown on (Table 3). Majority of the patients had PCV between 16 and 30. Most 41(82%) of them were underweight (Table 4). The prevalence of hypertension was 22%. (Table 5) shows frequency distribution table of their BMI and suggested that majority (82%) of them were underweight.

Table 1. Age distribution of 50 children with SCA

Age	Frequency	Percentage
<5	13	26
5-10	22	44
>10	15	30
Total	50	100

Table 2. Age, gender, blood pressure and anthropometric data

Parameters	SCA	Control	P-value
	Mean(SD)		
Age (years)	8.23(4.54)		
Gender [frequency (%)]			
Male	31(62%)		
Female	19(38%)		
Total	50		
Packed cell volume	22.48(4.1)		
Weight(kg)	27.75(13.75)		
Height (m ²)	1.27(0.29)		
Body mass index(kg/ m ²)	16.04(3.67)		
Systolic blood pressure	92.90(9.04)	98.08(10.73)	>0.05
Diastolic blood pressure	54.90(9.82)	62.64(7.57)	>0.05

Table 3. Packed cell volume distribution of children with SCA

PCV	Frequency	Percentage
≤15	2	4.0
16-20	16	32.0
21-25	20	40.0
26-30	10	20.0
≥31	2	4.0

Table 4. BMI distribution of children with SCA

BMI	Frequency	Percentage
<19	41	82.0
19-25	7	14.0
>25	2	4.0

Table 5. Correlation of clinico-laboratory parameters with the mean blood pressure in sickle cell anaemia patients

Factors	Mean(SD)	Mean(SD)	P-value
	Hypertensive	Non hypertensive	
Frequency (%)	11(22.0%)	39(78.0%)	
Age in years	6.56(4.16)	8.70(4.57)	>0.05
Packed cell volume	21.36(3.0)	22.79(4.34)	>0.05
Weight in kg	24.59(12.01)	28.64(14.22)	>0.05
Height in meters	1.22(0.29)	1.28(0.29)	>0.05
Body Mass Index	16.03(3.56)	16.04(2.75)	>0.05

4. DISCUSSION

Systolic and diastolic blood pressures in sickle cell anaemia patients from this study were lower than the control and were not significantly different from that of the controls. Moreso, lower blood pressure (BP) has been documented in children and adults with SCD [9-11]. The reasons for lower BP in sickle cell disease population are unclear but it may be partly attributed to the occurrence of increased renal tubular sodium and water excretion thus promoting lower arterial pressures [12-14]. Other reports however are of the opinion that the lower BP is due to a lower weight and increased vasodilation in adults and children with SCD [15,16]. With increasing age especially after adolescence, kidney function may deteriorate relatively faster in persons with SCD resulting in hypertension [16].

Children with SCD have classically been thought to have low normal blood pressures. In a study of 85 children, Aygun et al. [17] identified no hypertensive patients. In addition, in a Saudi Arabian cohort of 69 children with SCD aged 1–16 years old, blood pressure measurements were within normal range [18]. However, in the present study, 22% of patients had elevated blood pressures. Furthermore, a cohort study of thirty-eight children with SCD, based on in-clinic blood pressure screening showed prevalence of hypertension as 10.3% [19]. Another study found that BP was abnormal (hypertension and pre-hypertension) in 14.3% of the patients [20]. These findings coupled with our data suggest that hypertension may be under diagnosed in children with SCD when using standard clinic based assessments.

Most of the children with this condition in this study were underweight (low BMI). Studies have found an association between hypertension and BMI [20-23]. This may be due to the fact that patients with SCD have changes in plasma renin, endothelin and nitric oxide metabolites because of vaso-occlusion and those changes affect the balance between vasodilatation and vasoconstriction, which is not seen in undernourished children [23]. We did not confirm such association in our study.

5. CONCLUSION

There is no significant difference in the systolic blood pressure of children with sickle cell anaemia compared to age and sex matched controls. However, hypertension is well-defined,

prevalent, asymptomatic, chronic conditions in children with sickle cell anaemia. Based on the data in this study, this condition appears to be frequently undiagnosed by paediatric clinicians. Early, appropriate diagnosis is important so as to establish effective treatment for abnormal blood pressure.

CONSENT

All authors declare that 'written informed consent was obtained from the patient (or other approved parties) for publication of this study.

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

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