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Research Article

Prevalence of painful and anaemic crises among children with sickle cell anaemia in a tertiary hospital: A crosssectional study

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Abstract: Sickle cell anaemia (SCA) is one of the common causes of morbidity and mortality in children. Most of the hospital admissions among children with SCA in Nigeria are due to painful and anaemic crises. This study aimed to determine the prevalence of painful and anaemic crises among children with homozygous sickle cell disease attending tertiary hospital clinics. Two hundred and seventy-two children with SCA, aged 6 months to 15 years, were consecutively studied. It was a cross sectional study carried out at Ahmadu Bello University Teaching Hospital, Zaria, north-western Nigeria. Their age, sex and socio-economic status was determined through a structured questionnaire administered to the parents/ care-givers. Epi info statistical software was used in analysing the results. A p-value of ≤0.5 was considered statistically significant. Of the 272 children with SCA studied, 185 (68.0%) were in stable state while 87 (32.0%) had SCA crises. Their ages ranges from six (6) months to 15 years with a mean age (± 1 SD) of 6.4 ±3.8 years. Of the 87 subjects with SCA in crisis, 55 had painful crisis while 32 had anaemic crises giving a prevalence of 63.2% and 36.8% respectively. The type of SCA crisis was not affected by age, gender and social status. This study revealed a higher prevalence of painful than anaemic crises among children with SCA attending Ahmadu Bello University Teaching Hospital, Zaria and the type of SCA crises was not associated with age, gender and social class.
Keywords: children, painful crisis, prevalence, Sickle cell anaemia.

INTRODUCTION

Homozygous HbS disease (HbSS), an autosomal recessive disorder, was first described by Herrick in 1910 [1]. The prevalence rate of SCA among the black people of West and Central Africa descent is 2-3% [2]. Improved patient and/ or care giver education, multidisciplinary care and availability of advanced medical technologies have vastly improved the clinical outcomes of SCA in paediatric population. In high-income countries that provide neonatal diagnosis and care for patients, most survive well into adult life [3] and, because there is limited use of prenatal diagnosis[4,5], numbers of patients are rising steadily. Vaso-occlusive crises (VOC) contribute to frequent emergency department visits and hospitalizations in a subset of patients with sickle cell disease [6,7]. The purpose of this study was to determine the prevalence of painful crises among children with SCA attending a tertiary hospital. Children with SCA in crises are those with either painful or anaemic (hyper-haemolytic, aplastic, sequestration and megaloblastic) crisis defined as follows: Painful crisis – Recent onset of pains in the bones, hands, feet, abdomen and back with or without fever and bone tenderness. It also includes other conditions like acute chest syndrome which usually follows vaso-occlusion [8]. Anaemic crisis – These are patients with a significant drop of haemoglobin (greater than or equal to 2g/ dl) from their known value in steady state or patients presenting with haemoglobin of 4.5g/ dl or less where the steady state haemoglobin was not known [9].

The anaemic crisis is further subdivided into:

- Hyper-haemolytic crisis – Where there is evidence of a significant drop in haemoglobin > 2g/ dl in those whose haemoglobin had been estimated, with reticulocytosis 5 – 50%, jaundice and urobilinogenuria [10].
- Aplastic crisis: significant change in blood picture characterised by a fall in the haemoglobin level (>2 g/dl beyond steady state level) and reduced (<1%) or absent reticulocytes in the peripheral blood. The total white blood cell or platelet counts may or may not be affected. In addition, there is no significant increase in the unconjugated fraction of serum bilirubin [10].
- Acute sequestration crisis: significant change in blood picture characterised by a precipitous fall in the haemoglobin level and accompanied

by a rapidly enlarging spleen or liver (greater than 2 cm from the steady state level) and reticulocytosis above the steady state level for each individual patient. Signs of acute circulatory insufficiency such tachypnoea, tachycardia, and hypotension may or may not be present [10].

SUBJECTS AND METHODS

A total of 272 children with SCA (87 in crises and 185 in stable state) were recruited for this study. It was a cross sectional study carried out at Ahmadu Bello University Teaching Hospital, Zaria, north-western Nigeria. Subjects for this study included SCA children, both in stable state and in crises, who were recruited consecutively as they present to the hospital. Their ages ranged from 6 months to 15 years. The sex and socio-economic status (SES) of each subject was determined based on the method described by Ogunlesi [11].

Before patients are accepted into the clinic, their haemoglobin genotypes are confirmed with cellulose acetate haemoglobin electrophoreses and sickling tests. During the clinic visit, the type of SCA anaemia crises was determined through the history, physical examination and investigation. All subjects with recent onset of pains in the bones, hands, chest, abdomen, feet and back with or without fever and bone tenderness were considered to be having painful crisis. Subjects with a significant drop of haemoglobin (greater than or equal to 2g/ dl) from their known value in steady state or patients presenting with haemoglobin of 4.5g/ dl or less where the steady state haemoglobin was not known were considered to having anaemic crisis. Those with anaemic crisis were further classified into *hyper-haemolytic* (evidence of a significant drop in haemoglobin > 2g/ dl in those whose haemoglobin had been estimated, with reticulocytosis 5 – 50%, jaundice and urobilinogenuria), *aplastic* (significant change in blood picture characterised by a fall in the haemoglobin level, >2 g/dl beyond steady state level, and reduced (<1%) or absent reticulocytes in the peripheral blood) and *acute sequestration* (significant change in blood picture characterised by a precipitous fall in the haemoglobin level and accompanied by a rapidly enlarging spleen or liver, greater than 2 cm from the steady state level, and reticulocytosis above the steady state level for each individual patient) crisis. Ethical approval was obtained from the Ahmadu Bello University Teaching hospital's ethics committee.

RESULTS

Two hundred and seventy two (272) subjects with confirmed SCA, aged 6 months to 15 years were analysed. Their mean age (± 1 SD) was 6.4 ±3.8 years. Of the 272 subjects studied, 185 (68.0%) were in steady state and 87 (32.0%) were in SCA crisis. Of the 87 subjects with SCA in crisis, 55 had painful crisis while 32 had anaemic crises giving a prevalence of 63.2% and 36.8% respectively. The difference between those in painful and anaemic crisis is statistically significant (χ^2 = 12.8, p = 0.00). The mean age of SCA subjects in steady state was 6.8 ±3.9 years while that of those in crisis was 5.6 ±3.7 years and they were not significantly different from each other (χ^2 = 49.32, p = 0.10).

Table-1 showed the age and gender distribution of SCA subjects in crisis and in steady state. Although there were more males (57.4%) than females (42.6%) in the ratio of 1.3: 1 studied, the sex distribution in children in steady state was also not significantly different from that of children presenting in crises (χ^2 = 0.08, p = 0.39). There was no statistically significant gender difference between those sickle cell anaemia in crisis and those in steady state (p = 0.44). Table-1showed age and gender distribution of SCA subjects in crisis and in steady state.

Table-1: Age and gender distribution of SCA subjects in crisis and in steady state

	In steady state, n (%)			In crisis, n (%)		
Ages (Years)	Male	Female		Male	Female	Total
< 5	35 (33.3)	27 (33.8)		26 (51.0)	18 (50.0)	106 (84.1)
5-9	40 (38.1)	35 (43.8)		16 (31.4)	11 (30.6)	102 (71.5)
10-15	30 (28.6)	18 (22.5)		9 (17.6)	7 (19.4)	64 (44.3)
TOTAL	105(100.0)	80 (100.0)		51 (100.0)	36 (100.0)	272 100.0)

Forty-eight (17.6%) of the 272 subjects studied belonged to the middle socio-economic class families (class 3) while 88 (32.4%) and 136 (50.0%) belonged to the upper (classes I and II) and lower (classes IV and V) socio-economic class families respectively; Table II. Among the subjects in steady state, 30 (16.2%) belonged to the middle class families while 71 (38.4%) and 84 (45.4%) belonged to the higher and lower class families respectively. On the other hand, among the SCA subjects in crisis, 18 (20.7%) belonged to the middle class families while 17 (19.5%) and 52 (59.7%) belonged to the higher and lower class families respectively. The proportion of children with SCA in steady states in the upper socio-economic levels was significantly higher than the proportion of such children studied in crisis (χ^2 = 13.368, df = 4, p = 0.01); Table-2.

Table-2: Socio-economic classes of families of SCA subjects in steady state and in crisis

SEC	In steady state, n (%)		In crisis, n (%)	Total
I	14 (7.6)		6 (6.9)	20 (7.4)
II	57 (30.8)		11 (12.6)	68 (25.0)
III	30 (16.2)		18 (20.7)	48 (17.6)
IV	60 (32.4)		43 (49.4)	103 (37.9)
V	24 (13.0)		9 (10.3)	33 (12.1)
Total	185 (100)		87 (100)	272 (100)

Although there were more, 43 (49.4%), children with SCA in crisis, there was no difference between the age groups and the type of SCA crisis; χ^2 = 0.13, p = 0.94 (Table-3). There was no gender difference between children with SCA in painful crisis and those in anaemic crisis (p = 0.73). Although there were more children with SCA in crisis among the lower social class, there was no statistically significant difference between those in painful and amaemic crisis among the different age groups.

Table-3: Age, sex and social class distribution of SCA subjects in painful and anaemic crises

	Type of SCA crisis			
Age (years)	Anaemic crisis, n (%)	Painful crisis, n (%)	Total, n (%)	**P = value**
<5	15 (46.8)	28 (50.9)	43 (49.4)	
5-9	10 (31.3)	16 (29.1)	26 (29.9)	0.76
10-15	7 (21.9)	11 (20.0)	18 (20.7)	0.77
Sex				
Male	18 (56.3)	33 (60.0)	51 (58.6)	
Female	14 (43.7)	22 (40.0)	36 (41.4)	0.73
Social class				
Upper	6 (18.7)	11 (20.0)	17 (19.5)	
Middle	7 (21.9)	11 (20.0)	18 (20.7)	0.83
Lower	19 (59.4)	33 (60.0)	52 (59.8)	0.93
Total	32 (100.0)	55 (100.0)	87 (100.0)	

DISCUSSION

The prevalence rate of SCA painful crisis among the sickle cell anaemia children of 63.2% seen in this study is alarmingly high and worrisome considering the morbidities associated with this conditions. The finding of more children with SCA in painful crisis as compared to those in anaemic crisis found in this study indicates that painful crisis constitute most of the morbidities and hospital admissions associated with SCA. The hash weather conditions prevailing in the northern part of Nigeria where this study was carried out necessitating dehydration may explain the higher prevalence of painful crisis observed in this study as compared to the anaemic crisis. This finding of high prevalence of painful crisis is in keeping with the finding by Shapiro et al [12] in Philadelphia, the United States of America, where they reported painful crisis as a major cause of ongoing morbidity among subjects with SCA. Similar higher prevalence of painful crisis was documented by other studies in the United States [13] and in Great Britain [14], where they reported that painful crisis was the most common reason for emergency room and inpatient admission of patients with sickle cell anaemia.

On comparing the prevalence of painful crisis observed in this study and that of the UK study, the prevalence of painful crisis observed in this study was lower than that reported in Britain [14] children with SCA. The fact that many patients may not have presented to the hospital due to lack of awareness in our environment may have contributed to the higher prevalence of painful crisis observed in this study as compared to that observed in Britain where there is high standard of living and awareness. However, the prevalence of painful crisis observed in this study was higher than that observed in USA and Jamaica [12,15]. The high level of awareness and care available in the USA as compared to that found in Nigerian hospital may have contributed to the higher prevalence observed in our study.

On the other hand, the prevalence of anaemic crisis observed in this study was higher than the one observed in the Britain[14] study. It was also higher than that observed by Juwah *et al* [16] in Enugu. Malaria is an endemic disease in Nigeria and a leading cause of haemolysis in children with SCA [16]. This might explain the reason for the higher prevalence of anaemic crises in our study as compared to that reported in Britain where there is no malaria. High level of care rendered to children with SCA in Britain as compared to the care obtained in the locality where this study was conducted may also explain the difference in the

prevalence of anaemic crisis. Differences in the methodology and regional variation may have contributed to the differences in the prevalence observed between this study and the Enugu study. Because of the prevailing low socio-economic status and malaria endemnicity in our environment, one would have expected to find more children with SCA in crises among the younger age group and low social class. This study did not observe the contrary as age group, gender and socioeconomic status did not seemed to be associated with the type of SCA crisis.

CONCLUSION

There was relatively higher prevalence of painful than anaemic crises among children with SCA. Painful and anaemic crises remained the most common morbidities among children with SCA in our environment. There was no age, gender or social class difference between those children with SCA painful crises and those in anaemic crises.

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