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Original Research Article

Pattern of admission and outcome at a newly established sickle cell center in Nigeria

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ABSTRACT

Background: Sickle cell disease (SCD) encompasses a group of autosomal recessive haemoglobinopathies whose genetic mutation leads to the replacement of glutamic acid by valine in position 6 of the beta-globin chain. Most of the world's SCD burden is in Africa, where it is a major contributor to childhood morbidity and mortality. The study is aimed at determining the pattern of admission and outcome of patients seen at the sickle cell center Asaba, over a two-year period January 2022 to December 2023.
Methods: This was a retrospective descriptive study of children with SCD admitted into the sickle cell center of the Asaba specialist hospital. Information obtained included age, gender, diagnosis, month and year of admission, and outcome. Descriptive statistics was used to describe the demographic characteristics and chi-square to assess their association.
Results: A total of 338 children were seen, 34.5% of the patients were less than 5 years of age; males were 52.6% while 61.6% of the admission occurred during the rainy season. Vaso-occlusive crisis (80.6%) was the most common crisis seen; malaria (51.9%) was the prominent diagnosis made. Case fatality rate was 1.7%.
Conclusions: SCD s remains a major source of admission and mortality in our environment. A comprehensive SCD care plan is needed to reduce the adverse consequences associated with this disease.

Keywords: Sickle cell, Admission, Outcome

INTRODUCTION

Sickle cell disease (SCD) is an autosomal recessive genetic disorder caused by a single point mutation in the gene encoding the beta globin chain of haemoglobin.[1] It is often characterized by red blood cells becoming sickle-shaped and viscous thus clogging the blood flow especially in the small vessels resulting in sluggish flow, ischemia, pain and damage to the organs.[1,2] SCD is either inherited in the homozygous state also known as sickle cell anaemia (HbSS) or in a heterozygous form where an individual co-inherits HbS with another abnormal β globin gene, the commonest being SCD-SC, SCD-Sβ0 thalassemia, and SCD-Sβ+ thalassemia.[3]

SCD continue to be a major global public health concern. It is estimated that between 20 and 25 million people worldwide lives with SCD, of which 12-15 million lives in Africa.[4] Nigeria, the most populous black race in Africa has the largest number of HbSS patient in the world.[5] It is estimated that about 2-3% of the total 200 million Nigerian population has SCD, with an estimated 24% prevalence of sickle cell trait, 100,000 annual SCD birth and 100,000 annual SCD infant death.[6,7] This high mortality rate is influenced by poor access to health care,

poverty, ignorance and lack of comprehensive SCD management plan.

There is a wide variation in the clinical manifestations and severity of SCD and this probably due to the interaction of genetic and environmental factors such as infections, nutrition, geographic and climatic variation and socioeconomic status which has been shown to influence and modify the SCD patterns.[6,8,9] Children with SCD often suffer frequent hospitalizations due to various complications of the disease. Complications such as vaso-occlusive crisis, acute splenic sequestration, haemolytic crises, acute chest syndrome, stroke, pneumonia, malaria and severe infection have been reported as common reasons for admission in SCD children.[10-14]

There has been a significant improvement in the morbidity and mortality rates for children with SCD in high resource countries such as United States due to factors such as early diagnosis through newborn screening, prophylactic therapy, comprehensive care programs including hydroxyurea therapy and bone marrow transplantation. The above observation is different in poor resource countries like Nigeria. Mortality rate of 1.9% have been reported among SCD children in Ibadan.[15]

The main objective of this study was to describe the pattern of admission and outcome of children with HbSS at a newly established sickle cell center in Nigeria over a two year period. Findings from this study will guide future health care policies as there has not been any study on the morbidity pattern among sickle cell children in Asaba, Delta State, Nigeria.

METHODS

Study area

This was a retrospective descriptive study of children with SCD admitted into the sickle cell center of the Asaba specialist hospital. This center was established in 2021 and serves as a referral center for SCD in the state and environs. The pediatric section is headed by a consultant pediatrician with two resident doctors, two intern doctors, twelve nurses and other support staff. About 50 SCA patient are attended to weekly at the sickle cell center. A comprehensive paediatric cell care program is been offered at the Sickle cell center including newborn screening for SCD (HPLC assay), transcranial Doppler scan, automated exchange blood transfusion (EBT) using apheresis machine, counselling, provision of chemoprophylaxis, vaccines, hydroxyl-urea etc. Protocol for routine care includes: folic acid 5 mg, paludrine 100 mg, hydroxyurea 10-15 mg/kg/day, vitamin B complex, vitamin C 100 mg, tab penicillin V (Children <1 year 125 mg 2 times daily; 1 to 6 years: 250 mg 2 times daily, 6 to 12 years: 500 mg 2 times daily, 12 years and above: 1g 2 times daily.

Study population

The study population include all SCA children aged 0-18 years who were admitted in the sickle cell center of Asaba specialist hospital between January, 2022 and December, 2023.

Inclusion and exclusion criteria

All children aged 0-18 years who were diagnosed with SCA and managed within the period under review were included in this study. Children with other diagnosis other than SCA were not included in this study.

Study design

This is a retrospective descriptive hospital-based study of children with the SCD admitted into the sickle cell center of the Asaba specialist hospital Asaba, Delta State, Nigeria.

Data management

Data was extracted from the case notes of the patients seen during the study period. Data extracted included demographic variables, haemoglobin phenotype, type of crisis, associated infections and other complications of SCD, outcome of treatment and final diagnosis at discharge or death. Data were entered and analyzed with SPSS version 23.0. Categorical variables were presented as frequencies. Data presentation was done using frequency tables.

Definition of terms

Sickle cell was diagnosed using high performance liquid chromatography (HPLC). Diagnosis of sepsis was based on the presence of leucocytosis (WBC>20×10^9/L) or leucopenia (WBC<4×10^9/L) or thrombocytopaenia (<100×10^9/L) or neutrophilia or neutropenia (interpreted according to the age of the child).[11,12] Vaso-occlusive crisis was defined as an acute onset of pain in a child with SCD localized to the extremities, chest, back or abdomen.[9] Severe anaemia was defined as packed cell volume <15%.[11] Peripheral blood film (thick and thin smear) were used for the diagnosis of malaria. Rainy season in Nigeria is between the months of April to October.

Ethical considerations

Ethical clearance was obtained from the ethic committee of Asaba specialist hospital, Asaba and confidentiality was maintained throughout the study.

RESULTS

A total of 388 SCD patient were admitted into the SCD center over the period under review. The age of the subjects ranged from 5 months to 18 years with a mean

age of 9.5±5.7 years. Majority of the subjects (46.4%) were aged 10-18 years with more than 65% of the subject been aged 5 years and above. There were 204 (52.6%) male and 184 (47.4%) female given a male to female ration of 1.1:1. Most of the admission 239 (61.6%) occurred during rainy season. Nine patients died during period under review given a mortality rate of 1.7%. Most common cause of death was hyper haemolytic crisis. All mortality cases were aged >5 years (9-18 years).

Table 1: The demographics characteristics of the subjects.

Variables	N	Percentage (%)
Age (in years)		
0-5	133	34.3
6-10	75	19.3
10-18	180	46.4
Gender		
Female	184	47.4
Male	204	52.6
Season		
Rainy season	239	61.6
Dry season	149	38.6

Rainy season (April to October)

Table 2 shows the different form of sickle cell crisis among the subjects. Vaso-occlusive crisis (VOC) 191 (80.6%) was the most common form of crisis seen among the subjects. This was followed by hyper-hemolytic crisis 29 (12.2%) then acute chest syndrome 14 (5.9%). Vaso-occlusive crisis was commonest among male subject 101 (80.6%), and among those aged 16-18 years (89.7%). Acute chest syndrome was commonest among the male 8 (6.5%) and among children age 6-10 years (11.4%). Hyper hemolytic crisis was commonest among females 17 (14.9%) and among subject <5 years. Sequestration crisis occurred more in the male subject and 2 of the subjects were aged 11-15 years. Vaso-occlusive crisis, acute chest syndrome and hyper-hemolytic crisis occurred more during the rainy seasons while sequestration crisis occurred more during dry season.

Figure 1 showed the graphic representation of the admission pattern on monthly bases. The peak admission occurred in the month of May and June. Table 3 shows that the most common reason for admission was malaria 80 (51.9%) followed by sepsis 21 (13.6%) and pneumonia 20 (12.9%).

Table 2: Different sickle cell crises seen among the subjects.

Variables	VOC, N (%)	ACS, N (%)	HHA, N (%)	Sequestration, N (%)	Total, N (%)
Age (in years)					
0-5	47 (68.1)	6 (8.7)	15 (21.7)	1 (1.5)	69 (100)
6-10	35 (79.5)	5 (11.4)	4 (9.1)	0 (0.0)	44 (100)
11-18	109 (87.9)	3 (2.4)	10 (8.1)	2 (1.6)	124 (100)
Total	191 (80.6)	14 (5.9)	29 (12.2)	3 (1.3)	237 (100)
Gender					
Female	90 (78.9)	6 (5.3)	17 (14.9)	1 (0.9)	114 (100)
Male	101 (82.1)	8 (6.5)	12 (9.8)	2 (1.6)	123 (100)
Total	191 (80.6)	14 (5.9)	29 (12.2)	3 (1.3)	237 (100)
Seasons					
Dry	71 (75.5)	6 (6.4)	14 (14.9)	3 (3.2)	94 (100)
Rainy	120 (83.9)	8 (5.8)	15 (10.5)	0 (0.0)	143 (100)
Total	191 (80.6)	14 (5.9)	29 (12.2)	3 (1.3)	237 (100)

VOC: vaso-occlusive crisis, ACS: Acute chest syndrome, HHA: Hyper hemolytic anaemia

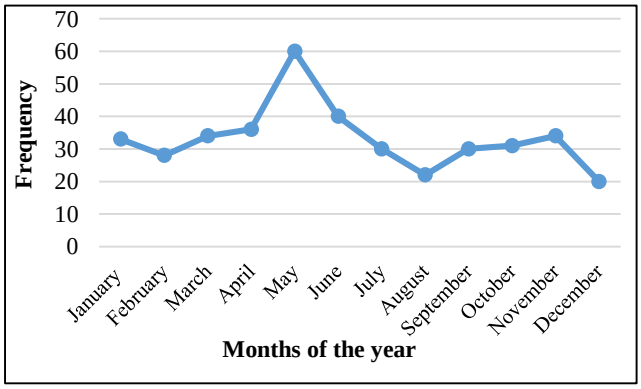

Figure 1: Monthly patterns of admission in sickle cell center.

Table 3: Diagnosis of patients at admission in sickle cell center (n=154).

Variables	N	Percentage (%)
Malaria	80	51.9
Sepsis	21	13.6
Pneumonia	20	12.9
Pharngotonsilitis	14	9.0
Meningitis	4	2.5
Stroke	4	2.5
Priapism	2	1.2
Hand-foot syndrome	1	0.6
UTI (Urinary tract infection)	2	1.2
Cellulitis	1	0.6
Diarrhea disease	5	3.2

DISCUSSION

This study shows that SCD still remain a major cause of admission and mortality in our environment. SCD in this study accounted for 10.4% (388/3229) of the total pediatrics admission. This finding is comparable to 10.1% prevalence reported in Zaria, Nigeria.[21] However, our finding is higher than 3.7% reported in Port Harcourt, 3.9% reported in Enugu and 4.1% reported in Uyo all in Nigeria.[19,25,26] The high prevalence recorded in this study could be attributed to the fact that the center is the sickle cell referral center for the whole of Delta State and other surrounding states. The massive campaign on the scourge of the disease by the state government and the role of state health insurance scheme in cushioning the financial cost has played a great role in early presentation to the hospital.

Male subjects predominated in this study and this finding is similar to what has been reported in other parts of Nigeria.[16,20,22] Though SCD is an autosomal recessive disorder and does not have gender predilection. However, some studies have postulated the role of nitric oxide in reducing the frequency of crisis among SCD and nitric oxide is more in female than male due to the protective effect of ovarian estrogen on Nitric oxide synthase and expression and this effect is considered to offer protective effects to females.[24]

These studies showed that children older than 5 years constitute 65.7% of the population. This finding is at variance to what has been previously reported in other studies in Nigeria.[19,21,22] This is an indication that most of our SCD patients are growing into adolescent and adult hood. This observation is similar to what has been reported in Jeddah, Saudi-Arabia.[1]

Most of the admissions 239 (61.6%) occurred during the raining season. This finding is similar to what has earlier been documented by other researchers.[19,22] The reason for this trend is not clear however, the effect of cold during the raining season could explain this occurrence. However, this finding is not in agreement with work done by Slovis et al in USA who did not see any seasonal variations in patients admitted with SCD.[23]

Vaso-occlusive crisis 191 (80.6%) is the most common form of sickle cell crises among the subject. This is not different to what has been documented in other studies.[16,19,20] Vaso-occlusive crisis was commoner among the male subjects possibly because they are more physically active than the females. Hyper haemolytic anemia 29 (12.2%), is the second most common form of crises seen among the subjects which also mirrors what Brown et al documented in Ibadan.[15] Hyper haemolytic crisis was more among the female subject than male subjects, the reason for this observation is not obvious.

The mortality rate in this study is 1.8% (7/388). This is similar to what was reported in Port-Harcourt 1.7% and 1.9% in Ibadan.[16,19] A mortality rate of 8.5% has also been documented.[20] The low mortality rate could be attributed to early diagnosis and comprehensive treatment protocol offered at the sickle cell center. In addition, the state health insurance program coves for most of the cost of treatment hence relieving the parents of significant financial burden if they were to pay out of pocket. Severe anaemia is the leading cause of death among the subject. This is attributed to default from routine clinic visits and late presentation to the hospital. This finding is not different to what was documented in Zaria, Nigeria, however CVA, adverse blood reaction and meningitis were the leading cause of death in Ibadan.[16,21]

Limitations

This is a single center-based study and might not give accurately the true burden of this illness.

CONCLUSION

SCD remains a major cause for admission and mortality among children in Delta state. Early diagnosis and prompt treatment often leads to a better outcome.

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Ethical approval: The study was approved by the Institutional Ethics Committee

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