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Clinical Profile of Children with Sickle Cell Anaemia in Nasarawa State, Nigeria: A Five – Year Review

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Abstract:

BACKGROUND: Sickle cell anaemia (SCA) is a chronic multi-systemic condition characterized by haemoglobin polymerization leading to erythrocytes rigidity, frequent haemolysis and vaso- occlusion. It is an autosomal recessive disorder associated with high morbidity and mortality. The prevalence of the disease varied from one location to the other.

AIM AND OBJECTIVES: This study determined the prevalence in Nasarawa state, the commonest crisis and the risk factors / complications of the disease.

MATERIALS AND METHODS: A hospital-based study of sickle cell anaemic patients seen between January 1st of 2013 to the December 31st of 2018. Hospital records, emergency units register and patients' folders served as the source of the secondary data. Analysis was done using SPSS version 23. Data was presented in tables, with categorical variables in frequencies and percentages while the continuous variables were presented as mean and stand deviation.

RESULTS: There are slightly more males than females with a ratio of 1.2:0.8. The mean age of diagnosing SCA was 9.7±6.6 years, with 55.7% of cases aged between 5 – 15 years. The prevalence of SCA in Nasarawa state was 0.02%. Two third of reasons for hospital presentation by sickle cell anaemic patients are due to vaso-occlusive crisis. About one in every five presents with deepening jaundice suggestive of hyperhemolytic crisis. Cellulitis and malaria were the commonest triggers leading to hospital admission in this study. Vaso-occlusive crisis warranting admission, while about a third had received blood transfusion at least once.

CONCLUSIONS: The burden of sickle cell anaemia is low in Nasarawa State. Vaso-occlusive crisis is the commonest acute manifestation at admission. Cellulitis and malaria are the leading triggers of such cases warranting hospital admission.

Keywords:

Burden, Nasarawa State, sickle cell anaemia

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Introduction

Sickle cell anaemia (SCA) is a chronic multi-systemic condition characterised by haemoglobin polymerisation leading to erythrocyte rigidity, frequent haemolysis and vaso-occlusion.[1] It is an autosomal recessive disorder occasioned by the production of abnormal haemoglobin S and it is associated with high morbidity and mortality.[2] Sickle cell disease occurs when there are two abnormal haemoglobin genotypes, i.e., having haemoglobin S with another abnormal haemoglobin such as C, D or thalassaemia, whereas the most common sickle cell disease genotype prevalent in people of African ancestry is HbSS.[3] Vaso-occlusive crisis (VOCs) and pain associated with such crises are hallmark symptoms in SCA and typically first manifesting in infancy.[4] These painful episodes can occur without warning and

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have been described as sharp, intense, stabbing and can be debilitating resulting in frequent emergency unit visits in the hospital.[5] Furthermore, they can be present with anaemic crisis of various forms as well as stroke with resultant major physiological, cognitive and emotional effects on the sufferers.[6]

Globally, there are 257,000 SCA patients, with many children born yearly with haemoglobinopathy, making it the most common haematological disorder.[7] SCA survival to adulthood in Africa was reported to be 10%–15% in the first decade of life, with a death rate of about 5% during subsequent decades.[7] An estimated 200,000 children with SCA are born in Africa each year, of which more than 50% of these births come from Nigeria.[7]

Saganuwan,[8] in 2016, found a 68.8% prevalence of sickle cell disease in North-western Nigeria.[8] A study by Raymond *et al*.,[9] in 2016, in the Akwanga Local Government Area of Nasarawa State revealed that more females had SCA compared to males.

Some of the common crises amongst sickle cell anaemic patients are as follows: Vaso-occlusive crisis also called painful crisis is the commonest crises found in this study, similar to earlier study.[8,10] This occur when tissue becomes deprived of blood flow from blockage caused by a sickled cell resulting in inflammation and extreme pain. Haemolytic crisis is another crisis which can be hyper-haemolytic and organ sequestration into the spleen (commonly) or the liver. Aplastic crisis, on the other hand, is a form of anaemic crisis characterised by an acute failure of erythropoiesis often following viral infections, especially parvovirus. Others include acute chest syndrome, with many of the above causing it. Blood transfusion, rehydration and use of analgesics are often necessary in order to preserve the patients' life and its quality.

Due to the paucity of data on SCA in Nasarawa State, despite it being an endemic environment like the rest of Nigeria, this study was conducted to determine retrospectively the burden of SCA, common SCA crises and complications of SCA in Nasarawa State, Nigeria, within the stated period.

Materials and Methods

Study area

Nasarawa State in North Central Nigeria is one of the 36 states in Nigeria, with an estimated population of about 2.6 million. The state has over 760 primary health centres, 17 general hospitals and two tertiary hospitals (Dalhatu Araf Specialist Hospital [DASH] Lafia and Federal Medical Centre Keffi), both serving as the state's referral centres.

The three health facilities selected for this study are DASH, General Hospital Akwanga (GHA) and Medical Centre Mararaba Gurku (MCMG). There are 18 state owned secondary / tertiary health facilities across the state. One health facility was selected from each of the three Senatorial zones through simple random sampling by balloting.

DASH is located in Lafia, the state capital, in the Southern senatorial zone with available bed space of 385 capacity with medically qualified consultants (six family physicians, five paediatricians, eight obstetricians, six internal medicine physicians, etc.) and more than two hundred nurses. GHA is located in the Northern senatorial zone with an available bed space of 80 capacity and having a clinical staff strength of 3 doctors, 3 pharmacists, 3 medical laboratory scientists and 33 nurses while that of Medical Centre Mararaba Gurku is located in the Western senatorial zone with 157 available bed spaces and having a clinical staff strength of 3 doctors, 4 pharmacists, 3 medical laboratory scientists and 44 nurses.

Study design

This was a hospital-based retrospective study of the admission records of patients diagnosed and treated for SCA from 1 January 2013 to 31 December 2018 at the DASH Lafia, GHA and Medical Centre Mararaba Gurku, respectively.

Study population

The study included all patients diagnosed and treated for sickle cell anaemia from 1 January 2013 to 31 December 2018 at the DASH Lafia, GHA and Medical Centre Mararaba Gurku in Nasarawa State.

Inclusion/exclusion criteria

All patients duly admitted with the diagnosis and treated for SCA from 1 January 2013 to 31 December 2018 in DASH Lafia, GHA and Medical Centre Mararaba whose records are complete were included in this study, while those whose folders could not be traced (six folders in all) were excluded from the study.

Sampling technique

One hospital in each of the three senatorial zones (as earlier explained) was chosen by simple random sampling. The hospitals (as stated above) in each of the senatorial zones were arranged in an alphabetical order, one each from each of the zones was randomly selected through balloting.

Data collection

Names and hospital numbers of all patients that were either seen at the clinics or at admission with a diagnosis of SCA were gotten from the clinic and admission

registers of doctors double checked with the Nursing record. These names with their corresponding hospital numbers, age and gender were taken to the medical records department for case note/folder retrieval.

A total of 179 patients were attended to within the stated period with the diagnosis of SCA-related crisis/ailments, with six of the folders missing. Of the six missing folders, one was seen but without the documents therein. Two research assistants who were youth corp members with background knowledge of computer, trained for a month and serving at the research unit under the supervision of the authors within the Epidemiology Department of the hospital.

Relevant data were extracted from the patient's folders using the questionnaire. Parameters assessed included age, sex, place of residence, occupation, common presenting complaints, crisis types and causes of admissions. SCA is diagnosed in the hospital using the haemoglobin electrophoresis conducted by one staff, an experienced medical laboratory scientist with master degree in haematological studies. He is being checked at interval by the head of the haematology unit of the laboratory for quality assurance.

The sickle cell anaemic patients were those already diagnosed and known clients on follow-up at the clinics (General Out-Patient Department, Medical Out-Patient Department, Surgical Out-Patient Department, Paediatric Out-Patient Department or the Ante-Natal Clinic) from where they are referred to the emergency units at the (Casualty, Emergency Paediatric Unit, Gynae Emergency and the Labour Room) for admission.

Data analysis

Data were analysed using SPSS (Statistical Package for the Social Sciences) version 23.0 (IBM SPSS Incorporated Chicago, Illinois, USA). Frequencies and percentages were computed for categorical variables, while means and standard deviation were determined for continuous variables that are normally distributed. Significant *P* value was at value less than 0.05. Obtained results were presented in tables.

Ethical considerations

Ethical approval was obtained from the Nasarawa State Ministry of Health Ethical Review Committee with Registration Number: NHREC/86/017. Confidentiality of information was treated with utmost regard and absolute trust.

Results

Socio-demographic distribution of the study population

A total of 83 (48.0%) SCA clients in this study were seen and managed at DASH. There are 96 (55.5%) males with a male-to-female ratio of 1.2:1. Most (153, 88.4%) of the participants were single, while 86 (49.7%) had primary school certificates with 15 (8.7%) having no any formal education. A total of 146 (84.4%) reside in the urban area and 114 (65.9%) were not gainfully employed at the time they were seen [Table 1].

Prevalence and age distribution of sickle cell anaemia in Nasarawa State

A total of 957,244 clients were attended to across the three selected health facilities in 5 years. DASH accounted for 644,324 (67.30%) of the total patients seen. Of these number (957,244), 173 (0.02%) were patients with SCA only. Majority (56, 31.40%) were aged between 5 and 10 years. The prevalence of SCA across the three health facilities in Nasarawa State was 0.02% [Table 2].

Common clinical manifestation of sickle cell anaemic patients at admission

In 75 (43.4%) of the cases in our study, bone pain was the reason for hospital visits. While 20 (11.6%) manifested with abdominal pain, 37 (21.4%) presented with yellowish discolouration of the eyes (jaundice)

Table 1: Socio-demographic distribution of the study population

Variables	*n* (%)
Centre	
DASH	83 (48.0)
GHA	51 (29.5)
MCMG	39 (22.5)
Sex	
Male	96 (55.5)
Female	77 (44.5)
Marital status	
Single	153 (88.4)
Married	20 (11.6)
Educational level	
None	15 (8.7)
Quranic	2 (1.2)
Primary	86 (49.7)
Secondary	62 (35.8)
Tertiary	8 (4.6)
Place of residence	
Rural	27 (15.6)
Urban	146 (84.4)
Occupation	
Unemployed	114 (65.9)
Civil servants	5 (2.9)
Business	1 (0.6)
Homemakers	1 (0.6)
Students	49 (28.3)
Artisan	3 (1.7)

DASH: Dalhatu Araf Specialist Hospital, GHA: General Hospital Akwanga, MCMG: Medical Centre Mararaba Gurku

Table 2: Prevalence and age distribution of sickle cell anaemia in Nasarawa State

Variables	*n* (%)
Total number of patients seen in the centres	
DASH	644,324 (67.30)
GHA	59,361 (6.20)
MCMG	253,559 (26.50)
Total	957,244 (100.00)
Prevalence of sickle cell anaemia	
DASH	117 (0.02)
GHA	11 (0.02)
MCMG	45 (0.02)
Total	173 (0.02)
Age groups (years)	
<5	38 (22.00)
5–10	56 (32.40)
11–15	42 (24.30)
16–20	16 (9.20)
>20	21 (12.10)
Total	173 (100.00)

Mean age±SD=9.7±6.6. SD: Standard deviation, DASH: Dalhatu Araf Specialist Hospital, GHA: General Hospital Akwanga, MCMG: Medical Centre Mararaba Gurku

Table 3: Common clinical manifestation of sickle cell anaemic patients at admission

Variable Presenting complaints	*n* (%)
Bone pain	75 (43.4)
Jaundice	37 (21.4)
Abdominal pain	20 (11.6)
Cough	13 (7.5)
Multiple (two or more complaints)	11 (6.4)
Back pain	6 (3.5)
Chest pain	3 (1.7)
Fever headache	2 (1.2)
Dysuria	2 (1.2)
Difficulty in breathing	2 (1.2)
Generalised body pain	1 (0.6)
Headache	1 (0.6)

and 11 (6.4%) presented with two or more clinical features [Table 3].

Causes of admissions, previous crisis and routine care

Cellulitis (71, 41.0%) and malaria (55, 31.8%) were the most common triggers leading to hospital admission in this study. Vaso-occlusive (painful) crisis warranting admission was seen in 100 (57.8%) cases; hyper-haemolytic crisis (29, 16.8%) was the most common of the anaemic crises, with mixed crises (vaso-occlusive and anaemic crises) in 26 (15.0%) cases. A total of 61 (35.3%) of the patients have had a blood transfusion at least once in the past [Table 4].

Discussion

This study found a low prevalence of sickle cell anaemia. Vaso – occlusive crisis was the commonest of the crises in these group of individuals with soft tissue infection, malaria and pneumonia the commonest predisposition to having crises. Blood transfusion is quite common amongst these patients.

The low burden of SCA found in this study could be due to the study design, wherein a retrospective study was conducted across three health facilities across the state. This did not afford us the opportunity to interact with the patients, carry out investigations and make a diagnosis ourselves. We relied on the diagnosis as written in the case notes. Two of the health facilities used were secondary health centres being run by younger doctors or medical officers with a limited knowledge base needed in differentiating between sickle cell diseases and SCA.

This is lower when compared with the 0.3% reported by Ejiofor *et al*.[11] in South-eastern Nigeria. Although, it is a retrospective study like the current study, the observed difference may be attributed to discrepancies in the diagnostic tool used. While their study population was newborn and screening done using iso-electric focusing machine, the current study was among patients aged six months and above using haemoglobin electrophoresis. It is much lower compared to the 3.5% reported by Adam *et al*.[4] in Sudan. The differences with our finding despite using haemoglobin electrophoresis like us may be because the Sudan study was a prospective cross-sectional study and done in a single hospital unlike ours.

Sack *et al*.[12] in Cameroon also found a higher prevalence of 1.27% compared to the present study. The variation with our study could be explained by the fact that Central Hospital Yaounde, where the study was done, is arguably the biggest public health facility better equipped with personnel and working tools, unlike the present study done in state health facilities, largely providing secondary healthcare.

We reported more males than females in this study, similar to the finding bySack and colleagues in Yaounde Cameroon.[12] But different from the report of a study by Olagunju in Ile-Ife.[13] The explanation could be that while our study is hospital based just like the Cameroon study, the study by Olagunju was community based and in South-western Nigeria.

Vaso-occlusive crisis was the most common acute manifestation found in subjects in the current study; this is understandable as most of our client's reasons for the presentation were due to pain from microvasculature obstruction by sickled red blood cell under hypoxic situations and leading to prostaglandin release and pains. This is similar to and reiterates the already known knowledge on the subject matter.[9,14]

Table 4: Causes of admissions, previous crisis and routine care

Variables	*n* (%)
Inter-current infections	
Cellulitis	71 (41.0)
Malaria	55 (31.8)
Pneumonia	27 (15.6)
Meningitis	8 (4.6)
Osteomyelitis	7 (4.1)
Typhoid	1 (0.6)
Others	4 (2.3)
Previous crisis	
Vaso-occlusive crisis	100 (57.8)
Hyper-haemolytic crisis	29 (16.8)
Mixed crises	26 (15.0)
Acute chest syndrome	13 (7.5)
Sequestration crisis	3 (1.7)
Aplastic crisis	2 (1.2)
Previous routine care	
Blood transfusion	61 (35.3)
Proguanil ingestion	57 (32.9)
Fansidar ingestion	24 (13.9)
Folic acid ingestion	107 (61.8)

Mean numbers of crisis±SD=1.4±0.9, mean frequencies of blood transfusion±SD=2.0±1.7. SD: Standard deviation

Skin infections manifesting as cellulitis, followed by malaria and pneumonia, were the most common infectious causes/triggers of the sickle cell anaemic patients presenting in crisis in this study. This is due to the increased susceptibility to malaria (unlike sickle cell trait) and other infectious agents, especially encapsulated organisms in this group of individuals.[15] The increased susceptibility can be attributed to many factors, namely auto-splenectomy or functional asplenia, reduced complement proteins, recurrent blood transfusion, hyperactivity of the bone marrow to constantly replenish the red cell lysis leading to the production of immature white blood cells called lazy leucocytes, etc.[16,17]

One out of every three of the patients in this study has had blood transfusion at least once in the past, this is also similar to earlier studies and it further predisposes them to infections.

This current study could, however, be limited by retrospective nature, limiting the diagnosis. The multiplicity of centres, as well as the method of laboratory diagnosis (haemoglobin electrophoresis), could be a limitation as only individuals older than 6 months could be reliably screened and diagnosed.

Conclusions

We found a low burden of SCA in this study. Vaso-occlusive crisis is the most common crisis, followed by hyper-haemolytic crisis (as a form of the anaemic crisis) amongst this cohort. Cellulitis, malaria and pneumonia were the leading trigger leading to crises that will warrant hospital admission. Most patients have had blood transfusion at least once in the past.

Recommendations

There is a need to introduce measures such as regular use of routine drugs and liberal fluid intake by sickle cell anaemic patients so as to minimise the propensity for painful (vaso-occlusive) crisis, which is the most common crisis in this study.

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Conflicts of interest

There are no conflicts of interest.

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