Original Article

Sickle Cell Anaemia: A survey of associated morbidities in Nigerian Children.

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ABSTRACT

AIM Sickle cell anaemia contributes the equivalent of 25% of under 5 deaths in Africa, with up to 16% of such deaths occurring in some West African countries. The aim of this study was to evaluate the prevalence and morbidities associated with sickle cell anaemia at the University of Port Harcourt Teaching Hospital (UPTH), Nigeria.

METHODS This was a retrospective review of case notes of children with sickle cell anaemia (aged 6 months to 18 years) that were seen in the Paediatric Haematology Clinic of UPTH, Port Harcourt, Nigeria from Jan 2009 to December 2009.
Data extracted from the patients case notes included age, gender, clinical features, genotype, history of blood transfusion, complications and crises.
Descriptive statistics were used to analyse data.

RESULTS There were a total of 169 children (aged 0.5-18 years) with sickle cell anaemia during this period. Of these, 92 (57.9%) were males while 77 (42.1%) were females giving a male/female ratio of 1.2:1. The mean age at diagnosis was 2.6 ± 2.1 years. Most 154 (91.1%) of the patients were under 10 years. More than two third of the patients had received blood transfusion ranging from one to seven times. Vaso-occlusive crises in the form of bone pains and hyperhaemolytic crises were the commonest crises while aplastic crises were not encountered. Commonly associated morbidities were malaria 58 (34.3%), dactylitis 43 (25.4%), pneumonia 18 (10.7%) and osteomyelitis 13 (7.1%).

CONCLUSION The study shows that the majority of patients with sickle cell anaemia were l0 years and below, pre supposing that many patients with sickle cell anaemia in our setting are dying in early childhood compared with more developed countries where these patients can live well into their 40s. There is therefore need for surveillance and education at the community level through the primary health-care system so as to increase public awareness of the problem and reduce morbidity and mortality of the affected individuals.

Keywords: Sickle cell anaemia; Survey; Morbidity; Children; Nigeria

INTRODUCTION

Sickle cell disease (SCD), first described by James Herrick in 1910 denotes all genotypes that contain at least one sickle gene in which hemoglobin S (HbS) makes up at least half of the haemoglobin(Hb) present. [1, 2] In addition to homozygotic HbSS (sickle cell anaemia), in which only HbS is produced, there

are at least 5 other major genotypes linked to the disease. [2]

The public health implication of SCD is enormous and is confounded by the unpredictable morbid states associated with the disease. Some patients with SCD may experience a mild disease course with minimal morbidity, while others repeatedly exhibit very severe symptoms.[3] Although the clinical course of SCD does not follow a pre-specified, uniform pattern, symptoms of chronic pain, acute anemia, infection, and other potentially debilitating complications are characteristic of the disease. Affected individuals present with a wide range of clinical problems that result from vascular obstruction and ischaemia.[3] When health impact is measured by under 5 mortality, sickle cell anaemia contributes the equivalent of 25% of under 5 deaths in Africa, with more than 9% of such deaths occurring in West Africa and up to 16% in some West African countries.[4]

Although an increasing number of children are surviving beyond 5 years there is still an increased risk of premature death.[4] A study done in Kano showed the highest prevalence in the 0-5 year age group and the lowest in the 21-25 year age group. There was no case found in the age group above 26 years.[5]

Sickle cell anaemia is thus a disease that needs to be thoroughly understood not only by clinicians, but also by the public in order to reduce its public health effects. Regular research on the disease and availability of information is therefore important to achieve this. The aim of this study was to evaluate the prevalence and associated morbidities of sickle cell anaemia at the University of Port Harcourt Teaching Hospital, Nigeria.

METHODS

This was a retrosprospective review of children with sickle cell anaemia (aged 6 months to 18 years) in the Paediatric Haematology Clinic of UPTH, Port Harcourt, Nigeria from January 2009 to December 2009. The Paediatric Haematology Clinic of UPTH operates once a week and receives referrals from other units within the Paediatric Department and its environs. All the patients were seen by the consultants. Only children with haemoglobin genotype SS established by the cellulose acetate haemoglobin electrophoresis were included for this study. Those with incomplete data were excluded.

Data extracted from the patients' case notes included age, gender, clinical features, genotype, history of blood transfusion, associated conditions and crises. Data was entered into a Microsoft Excel Spread sheet and analysed using descriptive statistics. Results were presented in tables.

RESULTS

There were a total of 6,072 children seen in the Paediatric Clinic of our hospital during this study period. Of these 169 children (aged 0.5-18 years) were confirmed to have sickle cell anaemia (SS) giving a prevalence of 2.9%. Among these children with SS, ninety two (57.9%) were males while 77 (42.1%) were females giving a male/female ratio of 1.2:1.The mean age at diagnosis was 2.6 ± 2.1years. Most 154 (91.1%) of the patients were under 10 years (Table 1). One hundred and two (60.4%) children had previous blood transfusions (ranging from one to seven). Vaso-occlusive crises in the form of bone pains (90%) and hyperhaemolytic crisis (60.4%) were the commonest crises encountered (Table 2). The clinical characteristics showed that 23 (13.6%) had bossing of skull bones, 98 (58%) jaundice, 152 (90%) clinical pallor, 32 (19%) splenomegaly. Malaria 58 (34.3%), dactylitis 43 (25.4%), pneumonia 18 (10.7%) and osteomyelitis13 (7.1%) were the most commonly associated conditions (Table 3).

Table 1: Age and Gender Distribution of the study population

Characteristic	Category	Number of patients (n=169)	Percentage
Gender	Male	92	57.9
	Female	77	42.1
Age (years)	<1	34	20.1
	1-<5	83	49.1
	5-<10	37	21.9
	≥10	15	8.9

Table 2: Types of Sickle Cell Crises

Crisis	No	Percentage
Bone pain	152	90
Abdominal pains	62	36.7
Acute chest syndrome	4	2.4
Hyper haemolytic	102	60.4
Sequestration	2	1.2

Table 3: Conditions Associated with Sickle Cell Anaemia

Conditions	No.	Percentage
Malaria	58	34.3
Osteomyelitis	13	7.7
Septic arthritis	7	4.1
Dactylitis	43	25.4
Pneumonia	18	10.7
Meningitis	5	3
Tuberculosis	3	1.8
Avascular necrosis of the hip bone	2	1.2
Cerobrovascular disease	3	1.8
Retroviral diseases	5	3
Epistasis	1	0.6
Priapism	2	1.2
Hyper-spleenism	6	3.6
Urinary tract infection	12	7.1

DISCUSSION

Sickle cell anaemia (SCA) causes serious agony to the individual, family and the society at large. Nigeria has the largest concentration of patients with sickle cell anaemia in the whole world. [2, 6] This study has demonstrated that 2.9% of cases seen in the Paediatic Clinic of our hospital were children with sickle cell anaemia. This is similar to previous African studies .[7,8] It is however at variance with a northern Nigerian study which reported a prevalence of 11.8%.[9] The fact that our study was a hospital based study could explain the differences observed . Males were more commonly affected than females. This observation is probably a reflection of the higher incidence of childhood illnesses in males in this environment. [10]

Also more than three quarters of the sickle cell anaemia patients in this study were below the age of 10 years. This can be explained by the fact that most children do not live beyond puberty as a result of frequent illness resulting in morbidity and mortality.[10] The most common presenting symptoms were pallor and jaundice. The sickle shape makes the cells stiff and sticky, causing them to become stuck in the vessels where they are destroyed by the spleen or simply haemolysed. Unlike normal red cells which last 120 days, sickle cells die after 10-20 days and because they cannot be replaced fast enough, the patients are chronically anaemic.[3] Jaundice is also a common symptom and sign of SCA. This is because the red cells haemolyse faster than the liver can filter bilirubin.[3]

We found bone pains in more than three quarters of our patients. This is as a result of microvascular occlusion of the bone marrow leading to ischaemic pain necessitating hospital admission and treatment with opioid analgesics.[11] Malaria was the most commonly observed infection among the patients studied. It is responsible for more than one million deaths annually. It modifies the clinical presentation of sickle cell disease and increases morbidity and mortality by precipitating vaso-occlusive and anaemic crises.[2, 12, 13]

Osteomyelitis was also a major presentation of SCA and accounted for 7.7% of cases in this study. This is however lower than 61.0% reported among Saudis [14] and 29.0% reported by Mijiyawa [15] in a neighboring West African country.

Priapism is a condition of painful, purposeless, persistent penile erection. We found a prevalence of 1.2% which is lower than previous studies. For instance, an earlier report [16] suggested that the prevalence of priapism was 2-6% . In a study of 52 men with sickle-cell disease, 20 (38%) reported having a history of priapism .[17] In another study, 27 of 98 (28%) reported a history of priapism on direct questioning .[18]

Acute vascular necrosis (AVN) complicating SCA has previously been reported to occur in 3-19% of

SCA patients.[14,15] We found a lower prevalence(1.2%). It is possible that most of the patients presented to traditional bone setters due to relatively high cost of orthodox medical care in Nigeria.

CONCLUSION

In view of the scale of the public health problem, a comprehensive approach to prevention and management of sickle-cell anaemia is urgently needed. Surveillance and education must be delivered at the community level through the primary health-care system so as to increase public awareness of the problem and reduce morbidity and mortality of the affected individuals.

FOOTNOTES

Conflicts of interest: The authors declare no competing conflicts of interest

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