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Original Research Article

The Pattern of Morbidity in Children with Sickle Cell Anaemia at the University of Uyo Teaching Hospital, Uyo, Akwa Ibom State, Nigeria

Eno-Obong Edet Utuk, Mkpouto Udeme Akpan

Department of Paediatrics, University of Uyo Teaching Hospital, Uyo. Akwa Ibom State.

Corresponding Author: Eno-Obong Edet Utuk

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ABSTRACT

Background: Children with sickle cell anaemia often require frequent and repeated hospitalizations and care due to the peculiar nature of this inherited genetic disease. There are few data on the causes of morbidity in Nigerian children with this disease, and no report so far, from this centre. Efforts at improving child survival and well-being due to diseases like this can only be effective if information about the common causes of illnesses and deaths are elucidated. This enables targeted focused interventions.

Aim: To document the morbidity pattern among children with sickle cell anaemia, managed at the University of Uyo Teaching Hospital, Uyo.

Methods: All children, six months to eighteen years with confirmed sickle cell anaemia seen and managed in the paediatric ward and children emergency unit over a two year period, from November 2012 to October 2014 were retrospectively studied for their pattern of morbidity.

Results: One hundred and eleven (4.1%) of a total 2686 patients were seen over the study period for various complications of sickle cell anaemia. The children were aged between six months and eighteen years with a median age of 6.5 years. Eighty-nine (80.2%) were males and twenty-two (19.8%) were females. There was one death during the study period. The leading causes of admission were malaria (72.1%), vaso-occlussive crises (70.3%), hyperhaemolytic crises (15.3%) and sepsis (13.5%). Other causes of morbidity included acute sequestration crisis (0.9%), dactylytis (4.5%), bronchopneumonia (1.8%), lobar pneumonia (1.8%), acute chest syndrome (1.8%), priapism (1.8%), chronic leg ulcer (1.8%), osteomyelitis (1.8%), septic arthritis (2.7%), hypersplenism (2.7%), nephrotic syndrome (1.8%), acute stroke syndrome (3.6%), avascular necrosis of the femoral head (1.8%), and haematuria in 0.9%. There was no case of aplastic crisis.

Conclusion: The major causes of morbidity in these children were vaso-occlusive crises and infections such as malaria. However, their spectrum of illnesses showed a multisystemic affectation. This multisystemic distribution of illnesses in affected children, further affirms the need for multi-disciplinary collaborations in their care.

Keywords: Sickle cell anaemia, morbidity, children, Uyo, Nigeria.

INTRODUCTION

Sickle cell anaemia is the commonest severe inherited disorder of humans. [1] The highest frequency of sickle-cell disease is found in tropical regions, particularly sub-Saharan Africa and the middle east. [2] Three-quarter of sickle-cell cases occur in Africa and a recent WHO report estimates a total of about 150,000 affected children born every year in Nigeria

alone. [1] The burden of non-communicable diseases like sickle-cell anaemia remains unacceptably high, particularly in sub-Saharan Africa, India and the Middle East. [2]

Many affected children suffer a lot of pain and illnesses. This could reduce their survival to adulthood. In 2010, there were about 29,000 deaths attributed to sickle-cell disease globally. [3] Those who do survive, consequently, often develop a lot of multi-systemic damage from the effects of chronic anaemia and haemolysis. [4-8] Sickle cell anaemia is a disease long known to have a variable clinical course. Efforts at improving child survival and well-being due to diseases like this can only be effective if information about the causes and trend of illnesses and deaths are elucidated, with focused intervention.

New approaches to management of this disease has improved diagnosis and supportive care over the last few decades, but many patients still have severe complications to contend with. [4-9] In many sub-Saharan African settings, health care facilities are inadequate and access to good health resource and drugs is limited to the relatively few persons who are able to afford fee-based private practices. Many affected children therefore do not receive adequate health care. They consequently, suffer a lot of illnesses associated with the disease throughout childhood. [2,3] This affects their quality of life, and thus contributes significantly to childhood morbidity and deaths from non-communicable diseases. This poor survival of children with sickle-cell anaemia in resource poor regions further worsens the myths and stigma already associated with the disease. [9-12]

There is a need to improve child health indices, particularly, to bridge the wide gap of ensuring achievement of the Millenium Development Goal 5 of reducing childhood morbidity and mortality by the year, 2015. In developing nations, like Nigeria, patterns of morbidity and mortality are not well defined and implementation of preventive care is poor. Knowledge of the present situation as regards child morbidity pattern in sickle-cell anaemia from various centres would be useful guides in assessing performance and identifying key areas where attention should be focused. Reports from various centres identified infectious diseases and the different types of sickle-cell crises, as major causes of morbidity for children admitted and managed. [5-8] There has been no documentation from this centre which serves a wide range of children within the state and its environs. Data obtained will help identify significant illness trends and guide effective prioritization of interventions to retard or halt disease progression to chronic organ disorders or multi-systemic failures. It would also serve as a reference point for future studies, and may provide a guiding framework for policy makers and health care planners to re-assess existing national policies, and delivery strategies. The monitoring, care and follow-up of affected children improves their quality of life, as they grow into adulthood.

MATERIALS AND METHODS

The study was retrospectively conducted in the Paediatric ward and children emergency unit of the University of Uyo Teaching Hospital, Uyo, from November 2012 to October 2014. Uyo is a fast growing city, located in the south-south geo-political region of Nigeria. The Teaching Hospital is the only tertiary health-care facility located at about six kilometers from the centre of the city. It renders both basic and tertiary healthcare services. The Paediatric unit is manned by all cadres of doctors and allied health-care staff including Consultants, Senior and junior residents, house-officers, nurses, pharmacists, and laboratory scientists. It offers twenty-four hours clinical service to all children.

All the patients aged between six months and eighteen years with confirmed sickle-cell anaemia managed in the Paediatric ward and children emergency unit were studied. The following data were obtained from their records and documented in a proforma: age, gender, definitive diagnoses, duration and outcome of admission. All their records were analyzed from admission to discharge. They were managed according to standard management protocol and guidelines. Data generated were entered into the computer, and analysis carried out with the Microsoft excel. Descriptive statistics was used to present the data. Frequency tables and percentages were also used to present the results.

RESULTS

There were a total of one hundred and eleven (111) children aged six months to eighteen years managed for various complications of sickle-cell anaemia during the two year study period. Of these 89 (80.2%) were male and 22 (19.8%) were female giving a male/female ratio of 3.67:1. This is illustrated in table 1. A prevalence of 4.1% of sickle cell anemia related admissions was obtained, with median age at diagnosis as 5.5 years. Most, 76 (68.5%) of the patients were under ten years of age. Vaso-occlusive crisis in the form of bone pain was the commonest crisis seen, followed by hyperhaemolytic and acute sequestration crisis. Aplastic crises were not encountered. Table 2 shows the list of the commonly associated morbidities in sickle cell anaemia. It included malaria 80 (72.1%), septicaemia 15 (13.5%), pneumonias (broncho- and lobar) 5 (4.5%) and dactylylitis 8 (7.2%). Chronic leg ulcers and avascular necrosis of the femoral head were 1.8% respectively. The leading causes of morbidity according to age groups were as listed in table 3.

Significant causes of morbidity in all age groups were noted to be malarial illnesses followed by the vaso-occlusive (bone-pain) crises. Other less causes of morbidity varied among the different age groups. Whereas, hyperhaemolytic crisis and septicaemia were commoner in children less than ten years of age, morbidities such as priapism, avascular necrosis of the femoral head, chronic leg ulcer and chronic osteomyelitis were seen in the older children and adolescents. Malaria was also a significant cause of morbidity in the under-five age group.

Table 1: Age and gender distribution of study population

	Gender		
Age	Male (%)	Female (%)	Total (%)
<5 years	24 (21.6%)	13 (11.7%)	37 (33.3%)
5-<10 years	28 (25.2%)	11 (9.9%)	39 (35.1%)
10-<15 years	9 (8.1%)	16 (14.4%)	25 (22.5%)
≥ 15 years	6 (5.4%)	4 (3.6%)	10 (9.0%)
Total	67 (60.4%)	44 (39.6%)	111 (100.0%)

Table 2: Distribution and frequency of various diagnoses made

Diagnosis	N	(%)
Vaso-occlusive crises	78	(70.3%)
Hyperhaemolytic crises	22	(19.8%)
Acute sequestration crises	3	(2.7%)
Malaria	80	(72.1%)
Dactylitis	8	(7.2%)
Bronchopneumonia	2	(1.8%)
Acute chest syndrome	2	(1.9%)
Acute stroke syndrome	4	(3.6%)
Urinary tract infection	1	(0.9%)
Priapism	2	(1.9%)
Lobar pneumonia	3	(2.7%)
Septicaemia	15	(13.5%)
Chronic leg ulcer	2	(1.8%)
Chronic osteomyelitis	2	(1.8%)
Septic arthritis	2	(1.8%)
Haematuria	2	(1.8%)
Nephrotic syndrome	1	(0.9%)
Cellulitis	3	(2.7%)
*Most children had more than one diagnosis		

The median duration of hospitalization was 8.0 days, with a range between one day to sixteen days. The outcome of most admissions in this study was good, as most of the children managed were discharged to the follow-up clinic. There was one only death (0.9%) recorded during the study period, which was due to an acute episode of hyperhaemolytic crises. Outcome and duration of hospitalization is shown in table 4. The reasons for the three patients who left against medical advice,

were severe financial constraints in two of the patients with acute stroke syndrome, and caregiver's impatience to stay for full recovery, in a child with severe vaso-occlusive crisis. The frequency of hospitalizations among the patients is shown in table 5. A greater percentage (81.1%) of the children had at least one episode of hospitalization during the period, and a few with greater than two episodes. The only death recorded was a child that died during the third episode of hospitalization.

Table 3: Major causes of morbidity in subjects according to age groups

Diagnosis	<5 years	5 - < 10years	10 - < 15 years	≥ 15 years
Malaria	28 (75.7%)	26 (66.7%)	20 (80.0%)	6 (60.0%)
Vaso-occlusive crisis	26 (70.3%)	24 (61.5%)	20 (80.0%)	8 (80.0%)
Hyperhaemolytic crisis	12 (32.4%)	8 (20.5%)	-	2 (20.0%)
Septicaemia	11 (29.7%)	4 (10.3%)	-	-
Dactylytis	8 (21.6%)	-	-	-
Bronchopneumonia	2 (5.4%)	-	-	-
Acute chest syndrome	-	2 (5.1%)	-	-
Sequestration crisis	-	2 (7.7%)	-	-
Priapism	-	-	2 (8.0%)	-
Avascular necrosis of femoral head	-	-	2 (8.0%)	-
Lobar pneumonia	-	-	2 (8.0%)	-
Chronic leg ulcer	-	-	-	2 (20.0%)
Chronic osteomyelitis	-	-	-	2 (20.0%)

Lesser causes of morbidity in the subjects were acute stroke syndrome, cellulitis, haematuria, and nephrotic syndrome.

Table 4: Outcome and duration of care in the patients

Outcome	< 2 days	– 7 days	>7 days	Total
Discharged	7	76	24	107
LAMA	0	0	3	3
Death	1	-	-	1
Total	8	76	27	111

Table 5: Number of hospital admissions in children with sickle cell anaemia within the last two years

No of hospital admissions	N	%
Once	90	81.1%
Twice	5	4.5%
Thrice	10	9.0%
Four times or greater	6	5.4%
Total	111	100

DISCUSSION

Sickle cell anaemia with its related complications constituted 4.1% of total admissions in the study period. This is closely related to the 3.9%, seen in a recent study in Enugu, [11] but higher than the 2.9% observed in Port-harcourt [13] both cities also located in the southern region of Nigeria. The results from this study showed a wide range of systemic complications causing morbidities, occurring in these children with sickle-cell anaemia. The clinical manifestations of sickle-cell anaemia results from two key pathological processes: vaso-occlusion and haemolysis. Sickled cells, along with the non-sickled red blood cells, leucocytes and platelets form heterocellular aggregates, which adhere to the vascular endothelium, causing obstruction to the lumen of small blood vessels. [14-18] This micro-circulatory occlusion, leads to acute and chronic tissue ischaemia and infarction with multi-system effects, particularly in the bones, lungs, brain, kidney and spleen. It is responsible for the acute painful episodes, crises and many of the long term episodes seen in sickle-cell anaemia. [10,14-18]

The clinical course of affected children is typically associated with intermittent episodic events typically referred to as 'crisis', which was noted to be a significant cause for hospital visits and admissions in this study. This is similar to reports from other centres within Nigeria [11-13] and other countries. [6,7] In almost all age groups in this study, malaria was a significant cause of hospital admission, followed by vaso-occlusive crisis. The same trend observed by Brown et al [5] in Ibadan, south-west Nigeria, and Opara [13] in Port-harcourt, south-south Nigeria. This is not surprising, as malaria has long been recognized as one of the most common precipitants of crisis in sickle cell anaemia. [10] Malaria was also documented in other studies as the most common precipitating

cause of crisis in endemic countries. [15,16] It is thus a significant pathogen in sickle-cell anaemia. Long term malarial prophylaxis has been shown to reduce the incidence of severe anaemia, number of crises and hospital admissions, as well as reducing mortality. [9,10,17,18] The use of prophylactic anti-malarial drugs like proguanil is routinely practiced, but from observations in this study, it may be more beneficial to improve on preventive strategies such as the use of insecticide treated nets, especially in this endemic locality.

Infections were also a common cause of morbidity in the under-five age group.

A major problem in children with sickle cell disease is altered splenic function resulting to increased susceptibility to infections by encapsulated organisms.

Overwhelming sepsis can develop rapidly, with no obvious primary focus of infection in children with sickle-cell anaemia. [10] The risk is confined almost exclusively to children with a reported incidence of 5.8/100 in children less than three years of age, 1.1/100 in children 5-9 years and 0.6/100 in children over 10 years. [1,2,15] During infection with any pathogen, changes occur at a cellular level, which predisposes to crises. These infections can lead to a range of complications, which are not readily reversed simply by treating the infection. For this reason, prevention remains the key strategy in management. Interventions such as the penicillin prophylaxis and pneumococcal vaccinations routinely used in some settings in the last two decades have dramatically reduced mortality, especially in children. The recommendations continue to evolve. In addition, infections can have more non-specific effects on the host's physiological milieu, increasing the risk of sickling. Fever causes water loss from sweating. This, coupled with reduced oral fluid intake, vomiting and anorexia, consequently leads to dehydration, which triggers crises. [10,17,18]

In one Nigerian center, [14] hyperhaemolytic crisis was the most common type of crisis in children with sickle-cell anaemia, but was not the case in this study. The incidence of 19.8% of hyperhaemolytic crisis in this study was lower than the 60.4% seen by George in Port-harcourt. [13] This could be attributable to the better uptake of routine anti-malarials, improved knowledge in caregivers on the treatment of malaria using the artemisinin combination drugs, and prompt presentation to hospital. Aplastic crisis was not encountered in this study. This has also been reported by others to be rare. [5,8,13,14] Such morbidities like priapism, avascular necrosis of the femoral head, constituted a low percentage of the total admissions, comparable to the 1.2% respectively documented in Port-harcourt. [13]

A greater percentage of the children (81.1%) were admitted at least once in the two year period with a few admitted more than twice within same period. This was also observed by Brown et al in Ibadan. [5] Children who spent more than a seven day period on hospitalization would also suffer significant loss of school hours, as well as working hours for care givers. This long hospital stay could cause destabilization of family routines and dynamics and all depicts the extent of burden in affected children, families as well as communities.

It will be helpful to improve preventive strategies like community awareness, genetic counseling, awareness of predisposing factors to common morbidities like the vaso-occlusive (bone pain) crisis. The use of expert counselors who are trained, was suggested in one study as a most effective low cost, and low technology strategy to use in any country for reduction of the problems associated with sickle cell disorder. [12] Also malaria preventive strategies like periodic distribution of insecticide treated bed nets should be sustained. Vaccine uptake should be

encouraged in care givers who can afford the pneumococcal vaccines, while government and non-governmental organizations should be encouraged to subsidize the cost of this vaccine, to encourage a wider coverage rate and uptake in many, who cannot afford. These strategies would go a long way in reducing the malarial illnesses, infections, and invariably the frequency of admissions/ morbidity.

Identification of children at birth by newborn screening programmes, instituting early preventive care, providing prompt and effective treatment to acute illnesses and prophylaxis against infections all contribute significantly to an overall positive impact on survival and quality of life. Countries with a large sickle cell disease population and adequate resources have commenced this. [9] This would be beneficial in a country like Nigeria.

It is important to encourage regularity in follow-up of sickle-cell patients, even in the steady-state period. This helps in adequate monitoring and treatment of affected children, to prevent onset of chronic illnesses which is seen in especially the older children and adolescents. Reducing the frequency of crises, ill health and systemic complications improves the patient's quality of life and may help to delay long term organ failures ultimately extending life expectancy.

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