Incidence of Sickle Cell Anaemia among Children Attending Maryam Abacha Women and Children Hospital, Sokoto

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Authors’ contributions

This work was carried out in collaboration among all authors. Author SYL designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Author JS managed the analyses of the study. Author JI managed the literature searches. All authors read and approved the final manuscript.

ABSTRACT

Sickle Cell Anaemia is still considered the most common genetic disease worldwide, causing morbidity and mortality in Sub-Saharan Africa, Mediterranean areas, Middle East and India. Nigeria, being the most populous black nation in the world, bears its greatest burden in Sub-Saharan Africa. This study was conducted to determine the incidence of Sickle Cell Anaemia among children attending Maryam Abacha Women and Children Hospital, Sokoto. A total of one hundred (100) blood samples were examined for the disease. Out of the 100 children tested for the disease, (59%) were normal (HbAA), (35%) were carrier (HbAS) and (6%) were Sicklers (HbSS). The result based on gender showed that female has the highest percentage of the disease (5%) against male subjects with only (1%). A child between the age group 6-10 years has the highest rate of sickle cell anaemia (3%) while age group 11-15 years had the lowest rate of the infection. Improved knowledge regarding Sickle cell anaemia disease and its comprehensive care among Nigerian physicians will enhance quality of care for affected childrens and policy for regular genotype test by government and other stakeholders before marriage among Nigerians will help to prevent the disease.
Keywords: Incidence; sickle cell; anaemia; children; Sokoto.

1. INTRODUCTION

Sickle cell anemia is still considered the most common genetic disease worldwide, causing morbidity and mortality in Sub-Saharan Africa, Mediterranean areas, Middle East and India [1]. Nigeria, being the most populous Black Country in the world, bears its greatest burden in Africa [1]. Sickle cell disease (SCD) is a molecular and genetic disease of haemoglobin. The disease of haemoglobin, hemoglobinopathies, can be divided into two main groups, the structural variant- Hb^S, Hb^C and Hb^E and the disorder of synthesis which can be traced back to the action of a mutant gene [2]. The mutant haemoglobin Hb^S, Hb^C and Hb^E are associated with sickling disorder of man, the abnormal haemoglobin is less soluble than normal haemoglobin Hb^A and therefore tends to crystallize out resulting in deformation of cells which instead of being round shape become sickle shaped [2].

Sickle cell disease (SCD) is the most common life threatening genetic disorder worldwide [1]. Much is known about the basic pathophysiology of SCD and the benefits of early intervention, and mortality from SCD has decreased to <1% in the Western world with a life expectancy of 53-60 years [2,3]. However 80% of SCD cases are in Sub-Saharan Africa where it has been reported that 50-90% of children born with Hb^S will not reach their fifth birthday and SCD accounts for up to 20% of neonatal mortality [4,5]. In 2006, the World Health Organization declared SCD to be a problem of major public health significance and a burden that must be addressed if recent improvements in overall child survival are to be consolidated [6].

Despite growing prevalence worldwide, the burden on Sub-Saharan Africa is expected to increase to 88% of cases by 2050 [7,8]. No global data regarding the precise numbers of children born with SCD and their hemoglobinopathies profile exist because, in contrast to Western countries, newborn screening for SCD is not available in most low income countries with the highest predicted burdens. Nigeria is known to bear the highest burden of SCD in the world and therefore the country is in urgent need of policies for prevention and management of SCD [7,9].

In Nigeria recent small-scale epidemiological studies have shown not only a high frequency of Hb^S but high prevalence of other associated haemoglobin genetic mutations like β-Thalassaemia, prompting the need for larger epidemiological research [10,11]. As new potentially curative therapies emerge and growing evidence for prevention strategies builds, it is of paramount importance that the burden of SCD in resource-poor countries is recognized to direct interventions and improve child survival. This research is therefore aimed at determining the incidence of the disease among children attending Maryam Abacha Women and Children Hospital, Sokoto with a view to finding out data on the disease and possible way(s) of preventing the spread of the disease in Sokoto and Nigeria in general [8,12].

2. MATERIALS AND METHODS

2.1 Study Area

This study was conducted at Maryam Abacha Women and Children Hospital, Sokoto. Maryam Abacha Women and children Hospital is a government owned specialized service hospital in Sokoto South Local government Area Sokoto State, Nigeria. Sokoto State is one of the Northwestern States of Nigeria. It is located at 13°05’N 05°15’E and cover the extensive geographical area of about 25, 973 square kilometer with a total population of over four million people. Sokoto State shares border with Zamfara State, Kebbi State and Niger Republic. The state consists of 23 local government areas, with its headquarter in Sokoto and its people are predominantly Hausa-Fulani whose main occupation is farming and animal husbandry [8,13].

2.2 Study Population

The study population consists of one hundred children attending Maryam Abacha Women and Children Hospital, Sokoto.

2.3 Study Design

This is an observational cross section study aimed at determining the incidence of sickle cell anaemia among children attending Maryam Abacha women and children hospital, Sokoto. The research was conducted during the rainy season from March to November 2019. The procedure was explained to all participant and were each given the consent form to sign. Questionnaires were distributed to generate information on their bio-data [12].
2.4 Administration of Questionnaire

A structured questionnaire was administered to the one hundred children recruited into study. Respondents who could not read or write in the English language were interviewed in Hausa by the researcher. The questionnaire was designed to provide information such as full name, age and sex.

2.5 Sample Collection

Blood samples were collected randomly among children attending Maryam Abacha Women and Children Hospital, Sokoto, using EDTA container by the laboratory technologies of the hospital.

2.6 Laboratory Analysis of the Samples

70 g of Ethylene diamine tetra acetic acid and 5.0 mls of distilled water, were mixed gently using pasture pipette, each bottle was labeled corresponding to the date, sex, and age of each sample. Six samples were centrifuged at a time using centrifuging machine KA 1000 model at increasing speed to a maximum of 4000 rpm for 5 minutes. The supernatant (plasma) was discarded and the sediment (RBC) was suspended by shaking the tube gently for each sample for 1-2 seconds. A volume of 0.5ml working solution, 2.0 mls of KCN stock solution 12.5 g potassium cyanide diluted to 250mls with 4 litres of distilled water plus 0.3 mls of EDTA and 120 mls of distilled water was pipetted into each of the tubes containing the sediment (RBC) [14].

A pipetted was used for the application of an extremely small amount (less than half a drop) of each haemolysate sample to each of the 6 segments on the plate. The wet cellulose acetate strip immersed in buffer (pH 8.6) (7.36 g of deithy/barbituric acid + 41.2 g of sodium barbitane + 4 litres of distilled water) was removed and excess buffer was also removed by blotting the strip between two pieces of filter paper. Finally, the teeth of the Shandon multiplicator were placed on the buffer impregnated cellulose acetate strip along a line 1.5cm from one of the lateral margins on the strip [13].

This was done gently but firmly so that the haemolysetes was applied directly on the same line of origin without shaking. The cellulose acetate strip (containing haemolysete) was fixed on both shoulders of the electrophoresis chamber so that sample 1-6 were aligned paralleled with the shoulders. The chamber was covered, the power pack was switched on and the electrophoresis is then carried out for 15-30 minutes. The cellulose acetate paper was removed, allowed to dry and the different genotypes were identified from their movement or differentiation in electrophoresis. The result (genotype) of each sample was recorded [15].

3. RESULTS

Out of 100 children tested for sickle cell anaemia, the result showed that 59% were found to be

Table 1. Overall genotype percentage in the population given from September to October 2017

| Genotype | Normal | Total (%) | Carriers | Total (%) | Sicklers | Total (%) |
|----------|--------|-----------|----------|-----------|----------|-----------|
| AA       | 59     | 59        | AS       | 29        | SS       | 1         |
|          |        |           | AC       | 6         | SC       | 5         |

Table 2. Genotype distribution of male and female population

| Sex    | Normal | Total (%) | Carriers | Total (%) | Sicklers | Total (%) |
|--------|--------|-----------|----------|-----------|----------|-----------|
| Male   | AA     | 22        | AS       | 10        | SS       | 1         |
| Female | AA     | 78        | AS       | 19        | SC       | 5         |

Table 3. Age distribution of the sample genotype and their population

| Age     | Total | (%)  | AA   | (%)  | AS   | (%)  | AC   | (%)  | SS   | (%)  | SC   | (%)  | Total | (%)  |
|---------|-------|------|------|------|------|------|------|------|------|------|------|------|-------|------|
| 0-5     | 41    | 41   | 26   | 26   | 12   | 12   | 1    | 1    | 1    | 1    | 0    | 0    | 16    | 16   |
| 6-10    | 33    | 33   | 18   | 18   | 11   | 11   | 1    | 1    | 0    | 0    | 3    | 3    | 23    | 23   |
| 11-15   | 26    | 26   | 15   | 15   | 6    | 6    | 4    | 4    | 0    | 0    | 1    | 1    | 17    | 17   |
| Total   | 100   | 100  | 59   | 59   | 29   | 29   | 6    | 6    | 1    | 1    | 5    | 5    | 100   | 100  |
AA, 29% were found to be AS, 6% were found to be AC, 5% were SC while SS had the least prevalence 1% (sickle cell anemia).

4. DISCUSSION

SCD is estimated to be the sixth leading cause of death in children in Nigeria [16]. Although intervention programs have been implemented for several other conditions such as HIV/AIDS and malaria eradication, there are no established programs for SCD despite the substantial associated morbidity and mortality. To our knowledge this is the first community based study to quantify the prevalence and pattern of SCD in Maryam Abacha Women and Children Hospital, Sokoto.

Studies from developed countries have clearly demonstrated the role of early diagnosis in improving the quality of life and survival in SCD. Appropriate preventive measures can be instituted through antenatal testing or neonatal screening. Pre-conception screening has been widely advocated in Greek Cyprus and Greece with some degree of success. Odunvbun and colleagues in South-Western Nigeria confirmed the acceptability of newborn SCD diagnosis [10]. Similarly there was a high level of acceptability in Sokoto despite the state perceived conservative, religious profile.

It was found that AA has 59%, followed by AS 29%, AC 6%, SC 5% and SS has the lowest prevalence of 1%. Also the incidence with respect to gender/sex showed that female has the highest incidence of 78% compared to male with 22%. Subject with respect to age, the result showed that, age group 6-10 years has the highest rate of sickle cell anaemia 3% and age group 0-5 year has the lowest rate of 2% [17,6].

Although, the participants displayed a higher level of knowledge, still there is lack of knowledge of predisposing factors to sickle cell disease. Many (25%) did not know that children with sickle cell disease can drink penicillin daily to decrease the rate of serious infection. Improved health education and educational campaign are needed to increase knowledge by intervening media campaign to increase sickle cell disease awareness [16,18].

5. CONCLUSION

Sickle cell disease is a common disease among people, the disease was as a result of inheritance of certain abnormal haemoglobin gene from each parent which may happen as a result of excretion, worry, dehydration or cold, and it was found that, there is no difference on white blood cell and plasma cell when compared for a normal person but the only difference is in the red blood cell, that is the packed cell volume as a result of RBC being in a sickle shape lacks enough hemoglobin making the PCV to be below in range. Marriage counseling for prospective marriage partners is crucial in fight against the disease.

6. RECOMMENDATION

It is our recommendations that, the sickle cell anaemia is a serious health problem, thus the need for government and management of Maryam Abacha Women and Children Hospital, Sokoto, to conduct sensitization and enlightenments with a view to reduce the burden and incidence of the disease.

The following are the recommendations of this research to the people with sickle cell anaemia.

I. Regular health maintenance of people with sickle cell disease.

II. Proper nutrition, good hygiene and bed rest is necessary for people with sickle cell anaemia.

III. Protection against infection and avoidance of other stresses, all are important in maintaining good health and preventing complications.

IV. Regular visit to a physician or clinic that provide comprehensive care which is necessary to identity early change in the patience health and ensuring immediate treatment.

V. Genetic counseling is also necessary for people with sickle cell anaemia.

VI. Similarly, there is need for policy for regular genotype test by government and other stakeholders among every individual before marriage so as to find out the suitable partner, this will help to prevent the incidence of sickle cell anaemia disease among children in the study area, Nigeria and world at large since the disease is genetically distributed [19,20].

CONSENT

As per international standard or university standard, respondent' written consent has been collected and preserved by the author(s).
ETHICAL APPROVAL

An introduction letter to undertake the study was obtained from the Head of Department of Biological Sciences, Sokoto State University, Sokoto and submitted together with the research proposal to the management of Maryam Abacha women and children Hospital, Sokoto through the chief medical director of the hospital. Ethical clearance and permission was granted by the management of the hospital which served as official approval for the research.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Attach B. Human pathology. Ibadan University Press Nigeria. 2001;122-123.
2. Balgir RS. Interventions and prevention of hereditary hemolytic disorders in two ethnic communities of Sundargarh District of Orissa: An Experience from KAP Studies India. Online Journal of Health and Allied Sciences. 2010;9(3):433-435.
3. Booth C, Inusa B, Obaro SC. Infection in sickle cell disease: A review. International Journal of Infectious Diseases. 2010;14(1):222-226.
4. Boyd J, Watkins A, Price C, Fleming F, DeBaun M. Inadequate community knowledge about sickle cell disease among African-American women. Journal of National Medical Association. 2005;97(1):63-67.
5. Howard J, Oteng-Ntim E. The obstetric management of sickle cell disease. Best Practice Research Clinic Obstetric Gynecology. 2012;26:25-36.
6. World Health Organization. Sickle cell anaemia: Report by the secretariat (WHO Publication No. A59/9). Geneva, Switzerland: World Health Assembly; 2006.
7. Jandi JH. A textbook of Haematology Little Brown. 2005;27-33.
8. Khatib R, Rabah R, Sarnaik SA. The spleen in the sickling disorders: An update. Pediatric Radiology. 2009;39:17-22.
9. Mabien A, Labbe E, Herbert D, Haynes J. Nurses attitudes and practices in Sickle Cell pain management. Applied Nursing Research. 2001;14(4):187-192.
10. Marlowe K, Chicella M. Treatment of sickle cell pain. Pharmacology. 2002;22(4):484-491.
11. Monahan DF, Sands KJ, Neighbors M, Marek FJ, Green JC. Phipps medical-surgical nursing: Health and illness perspectives Europe, Middle East and Africa edition. 8th edition Elservier. St. Louis; 2007.
12. National Institutes of Health. The management of sickle cell disease. NIH National Heart Lung and Blood Institutes; 2002. (Retrieved June 8, 2009) Available:http://www.nhlbi.nih.gov/health/prof/blood/sickle/SC_mngt.pdf
13. Olutunji PO. Post graduate doctor Africa. 2003;25(3).
14. Jaffer ED, Amrallah KF, Ali MK, Muhammad AN, Hassan AR, Humood MZ. Adult sickle cell disease patients and attitude towards the prevention measures of sickle cell disease crisis; 2009. Available:http://www.academicjournal.org/jnrm@2009academicjournal) (Accessed on 8/7/11)
15. Oludare GO, Ogili MC. Knowledge, attitudes and practices of premarital counseling for sickle cell disease among Youth in Yaba Nigeria. African Journal of Reproductive Health. 2013;17(4):175-182.
16. Tsaras G, Owusu-Ansah A, Osusu BR, Amoateng AY. Complications associated with sickle cell trait: A brief narrative review. The American Journal of Medicine. 2009; 122:507-512.
17. Platt O. Hydroxyurea for the treatment of sickle cell anaemia. New England Journal of Medicine. 2008;358(13)1362-1369.
18. Vishinsky EP, Neumayr LD, Earle’s AN, Williams R, Lennette ET, Dean D, Nickerson B, Orriger E, McKenzie V, Bellevue R, Daeschner C, Manci EA. Causes and outcomes of the acute chest syndrome in
sickle cell disease. New England Journal of Medicine. 2000;342:1855-1865.

19. Verduzco LA, Nathan DG. Sickle cell disease and stroke blood. 2009;114(25):5117-5125.

20. Villers MS, Jamison MG, DeCastro LM, James AH. Morbidity associated with sickle cell disease in pregnancy. American Journal of Obstetric Gynecology. 2008;199:125.e1-e5.