Original Article

Prevalence of Sickle Cell Disease among Children Attending Plateau Specialist Hospital, Jos, Nigeria

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Abstract

Background: An estimate of 250,000 children are born annually with sickle cell disease (SCD) worldwide and 75%–85% of the affected children are born in Africa; where mortality rates for those under age 5 years range from 50% to 80%. **Objective:** The present study was conducted to estimate the prevalence of SCD among children in Plateau State Specialist Hospital (PSSH), Jos, Nigeria. **Methodology:** Ethical approval was obtained from the Health Research Ethics Committee of the Hospital. Secondary data on age, gender, and region from the case notes of infants, children and/or adolescents; who received medical care in PSSH from 2012 to 2014 were used. Data were analyzed using frequency tables and Chi-square statistics. **Results:** The findings revealed that the prevalence of SCD in PSSH, Jos from 2012 to 2014 was 26.9/1000 population of pediatric patients. There was a gradual increase in the prevalence rate from 25.8/1000 in 2012 to 26.8/1000 in 2013 and 28.1/1000 in 2014. However, the case fatality rate of SCD gradually decreased from 15.4% in 2012 to 11.1% in 2013 and 10.3% in 2014. Chi-square test shows that the prevalence of the disease in relation to sex, age, and residence was not statistically significant ($P > 0.05$). Even though the case fatality rate of the disease decreased, its prevalence increased during the study. **Conclusion:** Therefore, preventive measure for SCD such as premarital genetic screening and counseling should be emphasized, especially in the southern and central geopolitical zones of Plateau state, where the prevalence was found to be higher.

Keywords: Children, prevalence, sickle cell disease

Introduction

Sickle cell disease (SCD) is the most prevalent inherited blood disease in the World Health Organization's African Region.[1] It is a genetic disorder of great epidemiological, clinical, and public health relevance in developing countries. It is an irreversible, untreatable health problem responsible for increased morbidity and mortality of school children.[2]

SCD is a genetic blood disorder affecting red blood cells (RBCs), with high morbidity and mortality rates.[3] The disease occurs due to a mutation of the beta globin gene of hemoglobin (Hb). This will result in the substitution of glutamic amino acid for valine at position 6 of the beta chain, thereby producing an abnormal hemoglobin called Hb S, instead of normal hemoglobin Hb A.[4,5] On deoxygenation, sickle hemoglobin undergoes a change in conformation that promotes intracellular polymerization, which leads to an alteration of the normal biconcave erythrocyte disc into the distinctive and pathological crescent shape. The resulting hemolytic anemia manifests as recurrent vasoocclusion and organ damage that together cause significant morbidity and early mortality.[6]

Sickle cell anemia seems to have targeted a certain race.[7] About 98% of the affected people are African-American. Researchers believe that the reason why African-Americans are affected the most is because of the malaria epidemic that occurs in Africa. It is believed that long ago, people who were infected with

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malaria had developed the odd/abnormal formation of their RBCs. In later generations, people who carry that trait can develop the disease by becoming infected with malaria. Other races include Caucasian, Mediterranean, and Middle Eastern.[8]

Sickle cell anemia does not target a certain age group. Each age group is affected equally because the disease is hereditary. Both parents must have the sickle cell trait (SCT) if the offspring would inherit the disease. Along with the ages, genders are both equally affected by sickle cell anemia. Males and females are affected equally because the disease is autosomal.[8]

The United Nations General Assembly has recognized SCD as a global public health concern due to the morbidity and mortality caused by the disease and the significant social and economic impact that results.[9] SCD affects 20–25 million people globally,[10] of which 12–15 million live in Africa,[11] but those affected in the developed countries account for only 10% of the world's SCD patient population.[9]

An estimate of 250,000 children are born annually with sickle cell anemia worldwide,[4,5] and 75%–85% of the affected children are born in Africa, where mortality rates for those under the age of 5 years range from 50% to 80%.[9,11] The highest prevalence of SCT in Africa occurs between the latitudes of 15°N and 20°S where the prevalence ranges between 10% and 40% of the population.[12] It is also estimated that 240,000 children are born with SCD annually in sub-Saharan Africa.[13] However, there have been studies in Africa that show a significant decrease in infant mortality rate, ages 2–16 months, because of the SCT. This happened in predominant areas of malarial cases.[14]

Quinn *et al*.[15] examined deaths in a cohort of newborn in Dallas and demonstrated that the overall incidence rate of deaths per 100 patients decreased from 0.67 in children from the period of 1983–1990 to 0.15 in the 2000–2007. Yanni *et al*.[16] compared mortality rates for children using the multiple cause of death files. They compared mortality rates for children during the period 1983–1986 with rates during the period 1999–2002. They demonstrated that the relative rate of mortality dropped – 68% for children 0–3 years of age, 39% for children 4–9 years of age, and 24% for children 10–14 years of age between the two time periods. In a study conducted in Lusaka, Zambia, Athale, and Chintu reported that the case-fatality rate among children with SS who were admitted to the University Teaching Hospital decreased from 18.6% in 1970 to 6.6% during 1987–1989.[17] Evidence of raised mortality has been reported in some studies,[18] but not in others.[19]

Despite effective ongoing comprehensive screening programmes, mortality from SCD is still high as indicated elsewhere in Brasil.[20,21]

Lanzkron *et al*.,[17] investigated mortality rates for children and adults with SCD in the United States and observed that mortality rate for adults appear to have increased during the same time period. It seems unlikely that this increase is due merely to an influx of younger patients surviving to adulthood and may reflect a lack of access to high-quality care for adults with SCD.[22]

Few studies[23-25] have been conducted to determine the prevalence of sickle cell anemia in children in some part of Nigeria, where sickle cell disorders are very rampant. However, prevalence study for Plateau is lacking. Therefore, the present study was conducted to estimate the prevalence of sickle cell anemia among children attending Plateau State Specialist Hospital (PSSH), Jos.

Methodology

The study was conducted in PSSH Jos, Nigeria. Secondary data from the case notes of infants, children and/or adolescents; who received medical care in PSSH from 2012 to 2014, were retrieved and analyzed.

Demographic data (age, sex, and regions) on confirmed cases of SCD were collected from the medical records departments for 3 years (2012–2014) from PSSH. Past records of SCD where available were thoroughly checked and recorded in excel spreadsheets.

Data collected were thoroughly cleaned for errors, completeness, and consistency checks. Information collected were entered into excel spreadsheet for storage and were later fed into Statistical package used for the analysis.

Inclusion and exclusion criteria

Inclusion criteria used in this study included individuals who were permanent residents in the study area. Exclusion criteria included individuals who were not permanent residents or on referral from LGAs other than the one under consideration.

Ethics consideration

This protocol received approval from the Health Research Ethics Committee (PSSH/ADM/ETH. CO/2015/004) PSSH.

Limitation

The retrospective survey was based on routinely collected data on reported SCD cases from the hospital. Due to the fact that retrospective data was used, its accuracy and completeness could not be fully verified. However, personnel at the hospital were well trained on diagnosis of SCD cases.

Results

The prevalence of SCD in PSSH from 2012 to 2014 was 26.9/1000 population of pediatric patients. There was a gradual increase in the prevalence rate from 25.8/1000 in 2012 to 26.8/1000 in 2013 and 28.1/1000 in 2014. However, the case fatality rate of SCD gradually decreased from 15.4% in 2012 to 11.1% in 2013 and 10.3% in 2014 as indicated in Table 1.

There was an even distribution of SCD across gender, (48.8% in males and 51.2% in females) as indicated in Table 2. Further, the calculated Chi-square confirmed that, there was no significant difference in the prevalence of SCD among male and female

children in PSSH, Jos (χ^2 = 0.1766, degree of freedom = 2, critical value = 5.99, P = 0.925 at the significant level of 0.05).

SCD was more prevalent among infants, toddlers and preschool children, (45.1%) followed by school age children, (32.2%) while it was less common among adolescents (24.4%) Table 3. However, the differences in the prevalence of SCD across these various developmental stages of children was not statistically significant, (χ^2 = 0.7183, degree of freedom = 4, critical value = 9.49 P = 0.925 at the significant level of 0.05).

The majority of SCD cases were from central Plateau, (32.9%) followed by Southern Plateau, (29.3%). 22.0% of cases were from Northern Plateau; while the least (15.9%) were from outside Plateau state as shown in Table 4. However, the distribution of SCD across these various geopolitical zones was not statistically significant. (χ^2 = 1.2265, degree of freedom = 6, critical value = 12.59 P = 0.97 at the significant level of 0.05).

Table 1: Sickle cell disease cases from 2012-2014 (n=3047)

Year	Total number of children treated in PSSH	Number of SCD cases	Prevalence of SCD per 1000 children	CFR of SCD (%)
2012	1009	26	25.8	4 (15.4)
2013	1007	27	26.8	3 (11.1)
2014	1031	29	28.1	3 (10.3)
Total	3047	82	26.9	9 (11.0)

PSSH: Plateau State Specialist Hospital, SCD: Sickle cell disease, CFR: Case fatality rate

Table 2: Sickle disease across gender

Year	Male	Female	Total
2012	12	14	26
2013	14	13	27
2014	14	15	29
Total (%)	40 (48.8)	42 (51.2)	82

χ^2=0.1766, degree of freedom=2, critical value=5.99, P=0.925 at the significant level of 0.05

Table 3: Sickle cell cases across age groups

Year	0-6 years	7-12 years	13-18 years	Total
2012	12	9	7	26
2013	11	10	6	27
2014	14	8	7	29
Total (%)	37 (45.1)	27 (32.9)	20 (24.4)	82

χ^2=0.7183, degree of freedom=4, critical value=9.49, P=0.925 at the significant level of 0.05

Table 4: Sickle cell disease across regions

Year	Northern plateau	Plateau central	Southern plateau	Outside plateau	Total
2012	6	8	7	5	26
2013	5	9	8	5	27
2014	7	10	9	3	29
Total (%)	18 (22.0)	27 (32.9)	24 (29.3)	13 (15.9)	82

χ^2=1.2265, degree of freedom=6, critical value=12.59, P=0.97 at the significant level of 0.05

Discussion

The prevalence of SCD in PSSH, Jos from 2012 to 2014 was 26.9/1000 population of pediatric patients. There was a gradual increase in the prevalence rate from 25.8/1000 in 2012 to 26.8/1000 in 2013 and 28.1/1000 in 2014. Similarly, Akinyanju[26] reported a high prevalence of sickle cell anemia in many Nigerian communities. This this was attributed to lack of premarital genetic counseling or couple going into marriage against medical advice when at risk of having children with SCD. The same could be true for the findings in the current study.

It was fortunate to note that the case fatality rate of SCD gradually decreased from 15.4% in 2012 to 11.1% in 2013 and 10.3% in 2014. This is consistent with the widely held view that interventions, such as penicillin prophylaxis and vaccination provided to people with SCD in early childhood, have played an important role in decreasing childhood mortality and preventing life-threatening infections.[15-17]

This study found that there was an even distribution of SCD across gender (48.8% in males and 51.2% in females). SCD affects males and females equally because the inheritance is "autosomal recessive." That is the affected gene is on one of the first 22 pairs of chromosomes that does not determine gender; hence, the disease occurs in children of carrier parents in the same proportion across gender in a ratio of 1:1 between male and female.[8]

In relation to age, the prevalence of the SCD was not statistically significant. This is because the disease does not target a certain age group. Each age group is affected equally since the disease is hereditary.[8] Even though the disease occurred more among younger children than adolescents; this was not significant; which concurs with the report of Akinyanju[26] that the prevalence of sickle cell anemia among babies born to Nigerian parents progressively decreases through late childhood, adolescence, and adulthood. This study finding may be due to the short lifespan of sickle cell children and the poor medical services in the developing countries like Nigeria; making most affected children to die before they reach adolescent age.

The SCD was more prevalent in the central and Southern Plateau (32.9% and 29.3%, respectively) as compared to Northern Plateau (22.0%). The reason is not known but it is likely to be attributed to the fact that the capital of Plateau state is located in the northern region; hence, people living in this area are likely to be influenced by their socioeconomic status, risk modifiers for instance SCD preventive measures such as premarital screening and behavioral patterns of the people. The complex interaction of these factors may have direct or indirect influence on regional SCD prevalence. This has been

reported for other diseases such as malaria among the people in this region.[27,28] The least prevalence of SCD (15.9% of case) was found among children outside Plateau state. This is likely to be due to distance from the hospital; as sickle cell children from other states are more likely to seek medical care in other hospitals located in their own respective states, rather than coming to PSSH.

Conclusion/Recommendations

We conclude that the prevalence of SCD in PSSH is increasing gradually, but the case fatality rate of the disease is decreasing gradually. Hence, there is need for preventive measures specific for SCD such as premarital genetic screening and public enlightenment campaigns and utilization of birth technology for couple who are already married. We also recommend that counseling should be emphasized, especially in the southern and central geopolitical zones of Plateau state, where the disease prevalence was higher.

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Conflicts of interest

There are no conflicts of interest.

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