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PREVALENCE OF SICKLE CELL DISORDER AMONG CHILDREN (0 – 10 YEARS) AT TWO TERTIARY HOSPITALS, ABIA STATE, NIGERIA.

Nursing

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ABSTRACT

Background: Sickle cell disorder as an inherited hemoglobin disorder, comprising sickle cell anemia (Hb SS) and some less prevalent but related conditions such as sickle hemoglobin C disorder (Hb SC) and sickle beta thalassaemia (HbSBthal). The aim of this study was to assess the prevalence of sickle cell disorder among children (0-10years) between 2013 – 2017 in two hospitals. **Methodology:** A cross-sectional descriptive survey was adopted; the design required no manipulation of variables and was used to study the retrospective prevalence of SCD in children (aged 0 month to 10 years) within ten years (10). The data was collected from medical records of both hard (physical files) and soft (computer) data base of children who were sick and either admitted or outpatient clinic. **Result:** In the two hospitals, 8470 children visited the hospitals within 10years,132 were investigated of genotype thus: 22 children were AA, representing (0.3%), 7 children were AS, representing (0.1%) and 103 children were SS, representing (98.4%). The number of male that visited the hospitals were 4553 (98.7) and female 3812 (98.8), 103 children were diagnosed of SCD, and out of this, 58(1.3%) were male and 45(1.2%) were female. Age 10 years (10yrs) was found to be the year with the highest occurrence of SCD with 17 SCDs representing (3.4%) of the age 10 children. In both hospitals, 2017 was discovered to have recorded the highest occurrence of SCD within the research period, followed by 2014, 2013, 2016 while 2015 recorded the lowest occurrence. There was a significant association between age of diagnosis and the occurrence of SCD among Children (0 – 10years) .There was no significant association between occurrence of sickle cell disease and centre/hospital (X2=0.97; p=0.33).There was significant association between prevalence of sickle cell disease and year of presentation with the highest proportion (1.8%) of sticklers diagnosed in the year 2017 (X2=14.49; p=0.01). **Conclusion and recommendation:** Sickle cell is a disorder that families and affected children are ignorant of due to culture, improper health care, low awareness and other factors. It was observed that many parents are not aware of their genotype and the diagnosis of the disorder was known late. Therefore, government should recommend a routine genotype test for all the parents before procreation.

KEYWORDS

Children, Genotype, Prevalence, Sickle cell disorder

1. INTRODUCTION

Sickle Cell Disorder (SCD) is one of the commonest genetic and inherited health problems that affect millions of people worldwide. It affect the structure, function or production of hemoglobin with three quarters of the cases occur thus in Africa [1]. Sickle cell disorder as an inherited hemoglobin disorder, comprising sickle cell anemia (Hb SS) and some less prevalent but related conditions such as sickle hemoglobin C disorder (Hb SC) and sickle beta thalassaemia (HbSBthal)[2]. [3] submitted that among all the variants, Hemoglobin SS is the most common pathological hemoglobin variant worldwide and majority of children born with Sickle Cell Anemia (SCA) die before reaching five years of age and consequently, about 150,000 Nigerian children are born each year with sickle cell anemia (HbSS) [4]. [5] Estimated that 231,000 children born with HbSS in 1996, 84% were born in sub- Saharan Africa [6]. They are reported to have migrated to South Africa, African-Americans and African-West Indians [7]. In developed nations, 1 in every 500 North American, African descendants and 1 in every 1,000 to 1,400 North American Hispanic descendants is a SCD carrier. The prevalence invariably will be higher in developing countries (Genetics Home Reference, 2007). In Turkey, [8] reported a 0.05% rate of SCT (HbAS) in one partner and both partners respectively, while in Saudi Arabia, [3] reported a SCT prevalence rate of 4.27% and 0.26% in one and in both partners respectively. Nigeria being the African most populated nation has risk of higher SCD prevalence and Nigeria's large population has ensured that over 40 million Nigerians are healthy carriers of the S gene. [9] States that about 25% of adults throughout the country have the sickle cell trait, AS, while the Hb C trait is largely confined to the Yoruba people of southwestern Nigeria in whom it occurs in about 6% of the people. The author further stated that other variant hemoglobin's including beta thalassemia are rare, but alpha thalassemia occurs in 39% (32% with 3 alpha-goblin genes; 7% with 2 alpha-globin genes). [10] Submitted that of a total of 5.4 million expected live births in 1988, about 90,000 had SCD and 1.1 million have the trait, AS. Although, countries around Nigeria also have an S gene carrier frequency of about 1 in 4 of their populations. The disorder, which is passed from parent to children spread all over the world as a result of the migration processes, slave trade, poverty, conflicts, inadequate health care, myths, stigma, ignorance, poor availability of resources to the public health and welfare sectors and economic inflation are severely curtailing access to appropriate medical and social services [11]. The disorder becomes life-threatening when there is hemolytic crisis, spleen sequestration crisis, or aplastic crisis, and clinical phenotype of sickle cell anemia is severe with manifestations occurring very early in childhood and mean Hb level 7.6 g/dl with HbF 5.9%.These complications are relatively higher in rural areas were medical care to mothers and children are less practiced [12].

Their disastrous health profile only serves to fuel the myths and stigma associated with sickle cell disorder in many communities in Africa. There is therefore an urgent need to improve the survival and quality of life for affected children in this sub region not only for the sake of families with affected members, but also as a strategy for combating ignorance and stigmatization within the communities. In most of these countries, health care facilities are inadequate and access to good health care is limited to the relatively few individuals who are able to afford fee-based private practices. Following the introduction of sickle cell genetic counseling to Nigeria in 1986 [13], it became possible for the first time ever to provide basic holistic care to hundreds of affected persons and their families.

Highlights in the United Nation has called for global efforts to bring attention to assess the burden of sickle cell disease and how to reduce it in Africa where about 85% of children with sickle cell disease are born in 2010. The World Health Organization (WHO) regional office for Africa proposed a sickle cell disorder strategy in official recognition of the fact that this disease is an important cause of child mortality in many African countries [14]. The purpose of this study was to assess the prevalence of Sickle Cell Disorder among children (0-10years) between 2013 – 2017 at Federal Medical Center Umuahia and Abia State Specialist Hospital Amachara, Abia State, Nigeria.

2. Material and Methods

A cross-sectional descriptive survey design was used for the study which required no manipulation of variables. The study was conducted in two hospitals namely Federal Medical Center Umuahia (FMC) and Abia State Specialist hospital Amachara both in Abia State. FMC is cited in the capital city of Abia State of Nigeria, it is the highest hospital and the only federal hospital in Abia state, has units, wards, specialty areas including children emergency units, under age wards, emergency pediatric unit etc. Abia Specialist Hospital Amachara is a hospital owned by Abia State Government, it is cited out sketched of the capital city, has wards and units. It has children ward were they admit both emergency and non emergency cases.

The two hospitals applied preventive, curative and rehabilitative approach in health care. They have health records section where all the files of the children are kept for care continuity and record purposes. Though they have computer to save the information but not comprehensive, so researchers sort using the hard and soft copies of the files were information of the patients for authenticity

2.1 Participants: The population for the study was children (0-10 years) who visited the hospitals for healthcare services from January 2013 – December 2017 according to the hospital data / record.

2.2 Research Design: The research members and the medical recorders selected the files of children within the age range from the file shelve starting from the list year (2013) to the highest year (2017). The collection of data took about 6months with the use of check list from the admission and discharge register and files. The annual information on genotype of the children (0 - 10yrs) from January 2013 – December 2017 was collected. Total number of 8470 children visited the hospitals within 10 years, out of this, 132 were investigated of their genotype using clinico-laboratory parameters.

2.3 Procedure: The check list used to collect the data was designed by the research team and heard two parts, first is the child status and parent's status. The relevant variables were coded in an excel spread sheet format, they include: Age of the child, gender, genotype, diagnosis, and age at sickle cell diagnosis, status of the child and parents socio demography. The research team drafted the format based on the existing child's history and items seen in the file of the child and the topic under study. To test for reliability of this instrument, pre-test method using split half technique on fifty (50) folders of the families who brought their children in the same hospitals and after six months the data of the same folders were collected to check the consistency. A reliability index (r) of 0; 95 was calculated using Pearson product moment statistics. For the validity, experts from measurement and evaluation from department of education Abia State university Uturu, Nigeria, different supervisors, from Nnamdi Azikiwe University Akwa, Anambra State, Department of Nursing Sciences, ethical committee members from the two studied hospitals, the heads of health record from the two hospitals. The ethical committee of the two hospitals approved the study before commencement. A copy of the approval latter (Ethical Clearance) was sent to the Head of Pediatric Department for approval and health records Department of the hospitals for the data collection. The health records collected the data; they refused the trained assistance in order to maintain confidentiality.

2.4 Data Analysis: There was no sample size calculated, according to [15], sample size calculation in retrospective study is not always mandatory. He said you can provide a time frame for your study as you are taking all the admitted patients in that hospital. The data was analyzed using the Statically Package for Social Sciences version 20 SPSS statistical package. Continuous variables were described with proportions when comparing groups of subjects with the use of frequencies, mean, tables, figures and charts., X (chi-square) test was applied to determine the significance difference observed and co-relation coefficient was used to analyze the hypothesis with Significant at $p<0.05$. Abia State Specialist Hospital had 209 as frequency of the participants with 2.5 % of the total participants, 208(99.5%) non sicklers 1 (0.5%) sicklers, while Federal Medical Center had 8159(98.8%) non sicklers, 102(1.2%), sicklers both had of mean different 0.97 (X) and 0.33 p-value of the disorder.

3. RESULT

Demographics

A total number of 8470 children visited the hospitals within 10 years (2013-2017), the year 2017 had the highest number of children that visited the hospital, the frequency of male children were 54,5% and female 45.5%. The death rate of the participant was not captured. Table 1 presented the socio-demographic variable. 132 were investigated of genotype thus: 22 children were AA, representing (0.3%), 7 children were AS, representing (0.1%) and 103 children were SS, representing (98.4%). Age above 10 years (>10yrs) is found to be the years with the highest occurrence of SCD with the percentage of 20.39%, followed by age 10 years (10yrs) 16.5%.

Age above 10 years (>10yrs) is found to be the years with the highest occurrence of SCD with the percentage of 20.39%, followed by age 10 years (10yrs) 16.5%, age 1 year (1yr) with 11.65%. It was closely followed by age 3 years (3yrs) which had 10.68%, age 5 years (5yrs) with 8.738%, age 6 years (6yrs) with 6.796%, age 7 years (7yrs) followed with 5.883%. Age 4 years (4yrs) took leadership of the next row with 4.854%, ages 2 years (2yrs), 8 years (8yrs) and 9 years (9yrs) taking 3.883% each, lastly age less than 1 year (<1yr) occupying the bottom with 2.913% (figure 1).

The figure 3 shows the prevalence of sickle cell disease in the health facilities, 98.78% of children were non sicklers, while 1.22% were sicklers within 2013-2017.

There was no significant association between occurrence of sickle cell disease and each of centre/hospital (X^2=0.97; p=0.33) and gender of the children (X^2=0.17; p=0.92). However, there was significant association between occurrence of sickle cell disease and year of presentation with the highest proportion (1.8%) of sicklers diagnosed in the year 2017 (X^2=14.49; p=0.01). There was also significant association between occurrence of sickle cell disease and age of diagnosis (X^2=45.81; p<0.01) (Table 2).

4. DISCUSSION

Findings of this study demonstrated that in every 100 children that visited the two health facilities, at least 2 had sickle cell disease (SCD). This is far higher than the findings reported by Nigeria Center for Disease Control and Prevention, in which the prevalence rate was 20-30/1000 live births SDC annually in Nigeria [16]. About 4.5% of the world population is carriers of sickle cell and about 300,000 infants are born with a major heamoglobinopathy every year. In the same vein, [17] further stated that the red blood cells (Erythrocytes) may contain normal hemoglobin AA only, a mixture of A and S (Hb AS) or S only. WHO further added that, 60 million carriers of sickle cell and 120,000 sickle cell homozygous are added every year in the entire world, at birth rate of 25 per 1000 live birth, about 45 million carriers and about 15,000 infants are affected. The result further shows that incidence of SCD was highest in 2017 within this ten (10) years mostly at FMC due to lack of awareness of the parents genotype before procreation. This corroborates the submission of [18], sickle cell disease is an inherited disorder of the blood which among other things imposes a shorter life span on the red blood cells (erythrocytes) which carry oxygen around the body.

The results show that male children had higher SCD than the female between the ages of 10 years (10yrs). The results also credited relationship between lack of adequate health care, low immune system to the complications of the disorder e.g. loss of splenic function occurs by first year of life and 90% by sixth year of life. .

However, lack of awareness by parent's genotype before procreation had really caused high mortality rate due to Sickle cell disease. Anecdotal evidence which shows that some of the people in Abia State have history of sickle cell disorder, which the researcher suspected that it is on increase and the chance of transferring both the trait and the disorder to their off spring and sustaining the disorder in the areas from generation to generation is paramount. Therefore, hospitals, religious body and government needs to set up policy to make sure that parent undergo a checkup to know their genotype and

make it assessable to both the rural and urban.

5. CONCLUSION

Sickle cell disorder, the abnormal Red blood cells in children from 0-10 years usually die after only about 10-20 days unlike the normal cells that live up to 120 days, Sickled RBC's can become trapped within the blood vessels and thus interfere with normal blood flow. The research was on the prevalence of sickle cell disorder among the children 0-10 years, it was observed that the disorder there were delay in diagnosis of sickle in children due to ignorant, low awareness creation and lack of equipments. The assessment tools for the data collection did not capture death ratio and complications following sickle cell. It is also a step for further research on the management of the disorder hence the study shows increase in the disorder.

Disclosure

The authors declare that they have no competing interest

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Declarations

Ethics Approval and Consent to Participate

The survey was fully accepted and scientifically approved by the two hospitals (Federal Medical Center Umuahia (FMC) and Abia State Specialist Hospital Amachara) both in Abia State with reference number FMC/QEH/G.596/VOL.10/373

Competing interests

The authors declare that they have no competing interests

Table 1: Socio-demographic and clinical variables of the participants

Variables	Categories	Frequency	Percentage
Hospital	State Specialist	209	2.5
	Federal Medical Center	8261	97.5
Year of presentation At the hospital	2013	1801	21.3
	2014	2022	23.9
	2015	906	10.7
	2016	1390	16.4
	2017	2351	27.8
Gender of children	Male	4613	54.5
	Female	3857	45.5
Genotype	AA	22	0.3
	AS	7	0.1
	SS	103	1.2
	Not checked	8338	98.4
Child status	Dead	0	0.0
	Alive	8470	100.0
Gender of parent	Male	44	0.5
	Female	159	1.9
	Incomplete data	8267	97.6
Parent's occupation	Civil/private sector	58	0.7
	Trading	75	0.9
	Artisans	49	0.6
	Farming	9	0.1
	Incomplete data	8279	97.7
Parent Marital status	Single	5	0.1
	Married	184	2.2
	Divorced	3	0.0
	Widowed	5	0.1
	Incomplete data	8273	97.6
Parent's genotype	AA	23	0.3
	AS	2	0.0
	Incomplete data	8445	99.7

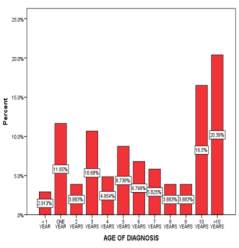

Figure 1: A bar chart showing the age of diagnosis of sickle cell disease among the children

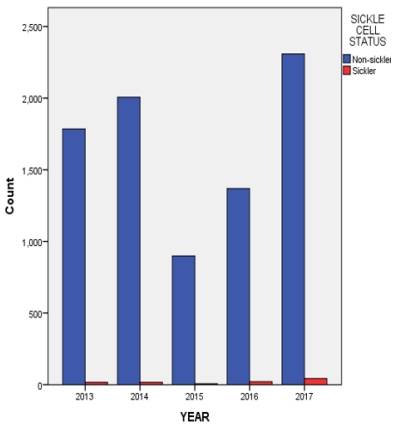

Figure 2: A chart showing the occurrence of sickle cell disease in different years of presentation at the hospitals

Year of Presentation	2013	16(0.9)%
	2014	16(0.8)%
	2015	7(0.8)%
	2016	21(1.5)%
	2017	43(1.8)%

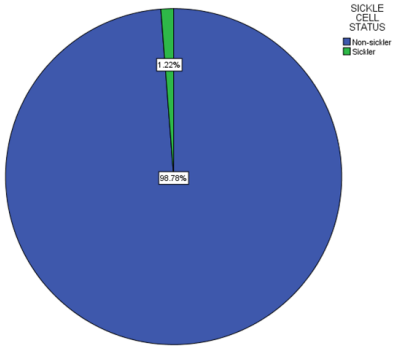

Figure 3: Pie chart showing the prevalence of sickle cell disease among the children in both hospitals.

Table 2: Chi-square test showing the association between occurrence of sickle cell disease and each of centres, gender, age and year of diagnosis

Variable	Categories	Sickle cell occurrence Non-Sickler	(n(%) Sickler	X^2	P
Hospital (Centre)	SSH	208(99.5)	1(0.5)	0.97	0.33
	FMC	8159(98.8)	102(1.2)		
Gender of Children	Male	4553(98.7)	58(1.3)	0.17	0.92
	female	3812(98.8)	45(1.2)		
Year of 2013 Presentation		1785(99.1)	16(0.9)	14.49	0.01*
	2014	2006(99.2)	16(0.8)		
	2015	899(99.2)	7(0.8)		
	2016	1369(98.5)	21(1.5)		
	2017	2308(98.2)	43(1.8)		
Age of Diagnosis	<1 year	1043(99.8)	2(0.2)	45.81	<0.01*
	1 year	1232(99.0)	12(1.0)		
	2 years	737(99.3)	5(0.7)		
	3 years	1050(99.2)	9(0.8)		
	4 years	431(98.9)	5(1.1)		
	5 years	1003(99.0)	10(1.0)		
	6 years	486(98.4)	8(1.6)		
	7 years	214(97.3)	6(2.7)		
	8 years	468(99.2)	4(0.8)		
	9 years	279(98.6)	4(1.4)		
	10 years	488(96.6)	17(3.4)		
	>10 years	935(97.8)	21(2.2)		

KEY:
*= Significant at $p<0.05$
SSH: State Specialist Hospital, FMC: Federal Medical Centre

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