Children living with Sickle Cell Disease (CLSCD): Socio-demographic profile of attendees in Federal Teaching Hospital, Gombe 2000-2014

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ABSTRACT

Background*: The sickle cell trait is highest in the North East region of the country with a prevalence of 32.6% and 27.9% in Borno and Yobe states respectively. Socio demographic circumstances of CLSCD have not been reported in the sub region.*

Objective: *To determine the socio-demographic characteristics of children living with sickle cell disease attending the Sickle cell Clinic of Federal Teaching Hospital, Gombe.*

Material and Methods: *Case notes of children diagnosed with sickle cell disease attending sickle cell clinic were retrieved and reviewed. Variables such as age, sex, tribe, religion, family history, Education level, occupation of parent and clinical events.*

Results*: 411 children with sickle cell anaemia were reviewed. 234(56.9%) were males and 43.1% (177) female. The Hb pattern was SS 86.2% (345), SF 12% (48) and SC 1.2% (5). Diagnosis of SCD was made between 2001 - 2005 in 64 (15.7%), 2006 - 2010 124 (30.4%) and 2011 - 2014 201(49.3%). 1.5% (6), 9.4% (37), 40.5% (159), 29% (114), 15.2% (60) and 4.3% (17) were between the ages < 6months, 7months1year, 15years, 610 years, 11-15 years and 16-18 years respectively. 88% (359) practiced Islam and 12% (49) Christianity. Of the ethnic groups, Fulani constituted 35.5% (146/411), Hausa 26.5% (109/411), Tera 6.3% (26/411), Tangale 4.4% (18/411), Bolawa 3.7% (15/411), Yoruba 1.7% (7/411) and Igbo 1.7% (7/411).*

Upper socioeconomic class (Class I and II) were 21.9% (90) and 10.2% (42); Middle socioeconomic class (Class III) were 89(21.7) and low socioeconomic Class (Class IV and V) were 9.7% (40) and 36.5% (150). 58.3% (240/411) and 39.1% (161/411) were from monogamous and polygamous families respectively. Consanguinity was found in 23.6% (97). 97.8% (402/411) of fathers and 96.8% (398/411) mothers had AS genotype. 56.9% (37/65) families reported 1 sibling death each from sickle cell anaemia; 20% (13), 3 deaths each; 18.4% (12), 2 deaths each and 1.5% (1), 5 deaths.

Conclusion: *Majority of children are from middle and lower level strata. Premarital screening should be strengthened.*

Key words: *Children, Sickle cell anaemia, sociodemography, FTH, Gombe.*

INTRODUCTION

Sickle cell disease (SCD) is one of the most common severe monogenic disorders worldwide.[1]It is a genetic disorder of great epidemiological, clinical, and public health relevance in developing countries. The disorder follows a more severe clinical course among patients in Africa than in those outside Africa.[2]

The World Health Organization recognized SCD as a global public health problem, as the overall number of babies born with SCD between 2010 and 2050 is estimated at about 14.24million.[2]

In the African region, majority of children with the most severe form of the disease (sickles cell anaemia) die before the age of 5, usually from an infection or severe blood loss.[3] In countries such as Cameroon, Republic of Congo, Gabon, Ghana and Nigeria the prevalence is between 20% to 30% while in some parts of Uganda it is as high as 45%.[3]

The global meta-estimate for the birth prevalence of homozygous sickle cell disease was 111.91 per 100,000 live births. There were however wide disparities by region, with a birth prevalence in Africa of 1125.49 per 100,000 compared with 43.12 per 100,000 in Europe.[4] The estimated number of new borns with SCA in Nigeria will be 140,000 by 2050 and country remains one of those most in need of policies for the prevention and management of SCA. [5]

Nigeria has the largest number of sickle cell anaemia (SCA) patients in the world with about 20 to 30% of the population trait carriers.[3] SCD affects about 2 to 3% of the Nigerian

population, this is the homozygous state. Borno and Yobe State has the largest number of sickle cell trait in Nigeria with prevalence of 27.9% and 32.6% respectively.[6]

Clinical features of SCD include acute pain episodes, anaemia, recurrent infections, and chronic end-organ damage.[7]

In Africa, significant morbidity and mortality are still associated with the disease, hence the need for effective and definite control measures.[89]

In Nigeria, efforts at public education on sickle cell disease and its prevention as well as carrier detection with genetic counseling have not made sufficient far reaching improvements in sickle cell disease control. There is still insufficient public knowledge about sickle cell disease.[10,11]

While some patients present with very severe clinical conditions, and are subject to many complications and frequent hospitalizations, others tend to show benign development, and some cases are almost asymptomatic. Both genetic and acquired factors contribute to this clinical variation. Among the acquired factors, the most important is the patient's socioeconomic conditions. [12]

Knowledge of the socioeconomic profile of SCD patients is essential to identify their needs, to contribute to improving resource allocation and to create and implement public health policies that benefit this population. [12, 13]Studies directed at these aspects of the disease are uncommon in Nigeria and in the sub region.

The objective of this study is to determine the socio-demographic characteristics of children living with sickle cell disease in Federal Teaching Hospital, Gombe.

MATERIALS AND METHODS

Study Area

Gombe is the capital of Gombe state. It is one of the six states that comprise North East Geopolitical zone in the country and one of the geopolitical zones with the highest levels of poverty and worse maternal and child health indices. [14]

Study setting

This study was conducted in Federal Teaching Hospital Gombe, a 500 bed hospital serving Gombe and neighboring states. The Federal Teaching Hospital, Gombe (FTHG) started providing services in the year 2000. It has emerged as a Centre for treatment, teaching and research in the sub region with large patient referrals. The FTHG is a regional Centre for sickle cell disease with capacity for neonatal diagnosis and social and genetic counseling. The center comprises of Paediatricians, Haematologists, Internal physicians, Pharmacists, Medical officers, Nurses, Laboratory Scientists, Counselors and Social workers, all trained at Sickle cell diseases foundation in Lagos, Nigeria. There are about 1000 patients enrolled in the center to date comprising of children and adults. Clinics are conducted regularly

Study population

Children aged 0-18 years diagnosed with sickle cell disease between 2000 to 2014 attending sickle cell clinic in Federal Teaching Hospital, Gombe.

Data collection

Records of children aged 0-18 years diagnosed with sickle cell disease attending sickle cell clinic in Federal Teaching Hospital, Gombe between 2000 and 2014 were retrieved. Variables such as age, sex, tribe, religion, family history, education level, occupation of parents and clinical events were retrieved and entered into structured questionnaire by the Paediatrics Data Management Team.

Data analysis

Records were imputed into Epi info Version 3.2 for analysis.

Ethical clearance

Clearance for this study was received from the Research and Ethics committee of the Federal Teaching Hospital Gombe.

RESULT

Four hundred and eleven children had sickle cell anaemia. Males were 56.9% (234/411) and 43.1% (177/411) were females. 10.9% (45), 40.6% (167), 28.9% (119), 15.1% (62) and 4.4% (18) were less than 1year, 15years, 610 years, 11-15 years and 16-18 years respectively (Table 1). Figure 1 shows the haemoglobin phenotype distribution amongst the study population with HbSS constituting 86.1% (354), HbSS+F 12% (49), and HbSC 1.2% (5) The diagnosis was made in 33.8% (139) of children at less than 1 year of age, 51.6% (212) 1-5 years; 7.8% (32) 6-9 years and 6.8% (28) 10-18 years.

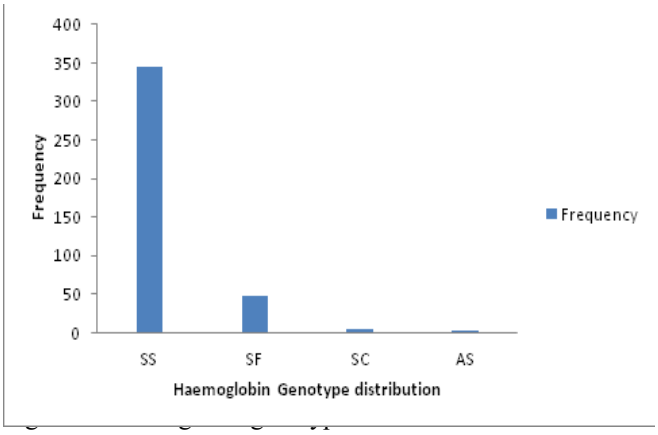
Figure 2 shows that 16.5% (68) of the children were diagnosed with sickle cell disease between 2000-2005; 31.9% (131), 2006-2010 and 51.6% (212), 2011-2014. Figure 3 shows that 88% (362) of children belong to Islamic faith while 12% (49) are Christian. Of the ethnic groups, Fulani constituted 35.5% (146/411), Hausa 26.5% (109/411), Tera 6.3% (26/411), Tangale 4.4% (18/411), Bolawa 3.7% (15/411), Yoruba 1.7% (7/411), Igbo 1.7% (7/411) and other ethnic groups 20.1%(83) (Figure 4).

The index subject was the first child in the family living with the disease in 20.2% (83/411) of cases, 2nd and 3rd child in 18.0% (74) of the families, 4th, 5th, 6th, 7th, 8th, 9th and 10th child in 11.0% (45), 7.8% (32), 10.7% (44), 4.9% (20), 3.2% (13), 2.1% (9) and 4.1% (17) respectively. The total number of children in a family was reported as 1-5 children in 55.5% (228) families, 6-10 children in 31.4% (129) and 13.1% (54) have more than 10 children. (Table 2)

Figure 5 shows the socioeconomic status of CLSCD using Oyedeji's classification.[15] Upper socioeconomic class (Class I and II) were 21.9% (90) and 10.2% (42); middle socioeconomic class (Class III) were 89(21.7) and low socioeconomic Class (Class IV and V) were 9.7% (40) and 36.5% (150). 58.3% (240) of the children were from monogamous, 39.1% (162) from polygamous families and 2.2% (9) from divorced families. 97.8% (402) of fathers and 96.8% (398) mothers had AS genotype. Consanguinity was reported in 23.6% (97) of marriages. 65 families reported child deaths from Sickle cell disease; 1 death each in 56.9% (37/65) of families, 2 in 18.4% (12), 3 in 20% (13), 4 in 2 and 5 deaths in 1 family.

Table 1: Age and Sex distribution of Children living with Sickle cell anaemia

	SEX			
Age group	Male (%)	Female (%)	Total (%)	P value
<1yr	30(66.7)	15(33.3)	45(10.9)	0.414
1-5yrs	88(52.7)	79(47.3)	167(40.6)	
6-10yrs	70(58.8)	49(41.2)	119(28.9)	
11-15yrs	34(54.8)	28(45.2)	62(15.1)	
16-18yrs	12(66.7)	6(33.3)	18(4.4)	
Total	234(56.9)	177(43.1)	411(100)	



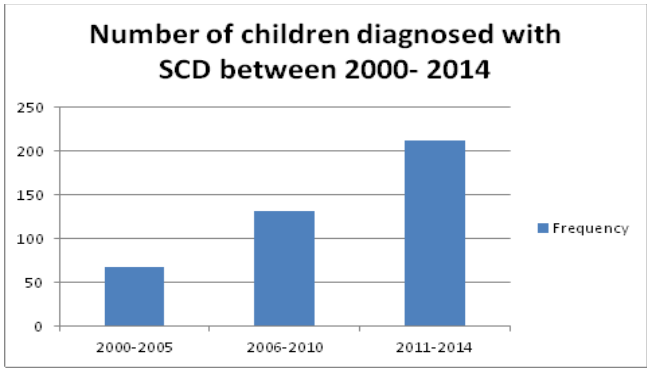


Figure 2: Number of children diagnosed with Sickle cell disease between 2000- 2014

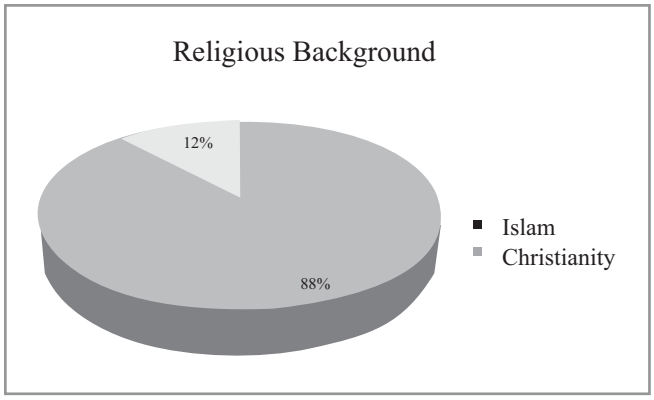


Figure 3: Religious background of children living with sickle cell

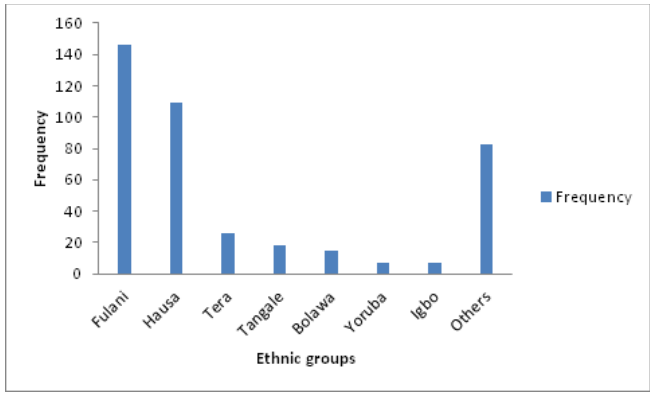


Figure 4: Ethnic background of children living with Sickle cell disease

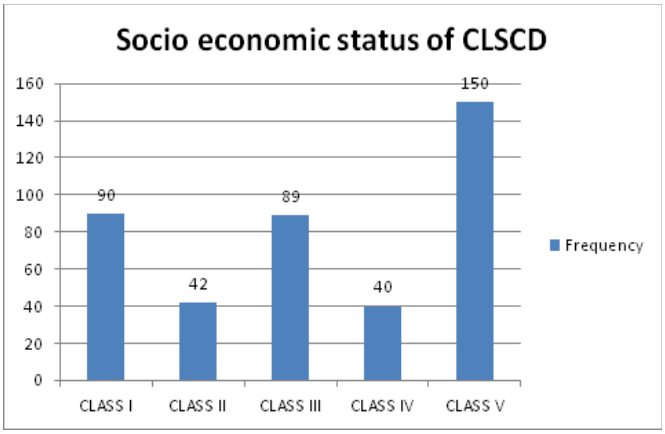


Figure 5: Socioeconomic Status of children living with sickle cell diseases

Table 2: Demographic characteristics of children with sickle cell disease

Demographic characteristics	n (%)
Age at diagnosis	
>1yr	139(33.8)
1yrs-5yrs	212(51.6)
6yrs-9yrs	32(7.8)
10yrs-18yrs	28(6.8)
Family size	
1-5	228(55.5)
6-10	129(31.4)
>10	54(13.1)
Birth order	
1st	83(20.2)
2nd	74(18.0)
3rd	74(18.0)
4th	45(11.0)
5th and beyond	135(32.8)
Marital Status of parent	
Monogamy	240(58.4)
Polygamy	162(39.4)
Divorced	9(2.2)

Table 3: Consanguinity among families with sickle cell disease

Consanguinity	Islam (%)	Christianity (%)	p-value
Yes	96(99.0)	1(1.0)	0.000
No	266(84.7)	48(15.3)	

DISCUSSION

To the best of our knowledge, this is the first report of the socio-demographic profile of children living with sickle cell disease in North-East Nigeria.

In our study there were more males than females attending Sickle cell clinic of our health facility. This gender difference is similar to reports from Maiduguri, [7] Lagos, [16] Jos, [17] Benin-City [18] and Ghana. [19]

Majority of children living with SCD in our study were in the under -5 age group. This is similar to the findings from Maiduguri, [7] Lagos, [16] Jos, [17] Enugu, [20] Sokoto [21] and Ife. [22] Adolescents constituted a fifth of children living with sickle cell disease in this study; this in contrast to 30% in Benin,[23] but similar to Jos [16] and Sokoto.[21] Adolescents with SCD were lower in numbers in Ife [22] and Enugu. [24] These differences were as a result of sample sizes and the various different age limits for inclusion as adolescents. Many babies born with sickle cell disease in Nigeria die before they reach puberty[25] with increased risk of death between 1 and 3 years of age in both low and high income settings.[8,9]

HbSS+F percentage in our study is slightly higher than9.6% and 9.9% reported in Ife by both Akinlosotu et al [26] and Adeodu et al [22] respectively. Our finding is much higher than Jiya et al (6.1%) [21], Isah et al (2.9%) [27] and Saganuwan et al (3.6%) [28] all from Sokoto and 6.5% by Dangana [29] et al from Abuja. Reports from India [30] and Uganda [31] shows result consistent with our finding. While test methods, age, sex, hydroxyurea, haplotype and geographic region can influence its distribution and levels, [22,26] the level of HbF continues to drop and stabilizes at age 5-10 years and HbF ameliorates the clinical and haemotological consequence of sickle cell disease. [26,32,33.]

Adegoke et al [23] in Ife reported HbSC of 9.6% in CLSCD in contrast to 1.2% in our study and 1.5% by Jiya et al [22] in Sokoto. Inusa et al [34] reported 0.13% prevalence of HbSC in children in three contiguous states of Kaduna, Katsina and Abuja. HbSC is a milder disease with less frequent and less severe symptoms. [32] [33] Nationally representative data through multi Centre surveillance are urgently needed to deepen the understanding of this disease.

One-third of our patients were diagnosed in the first year of life. This is comparable to findings by Ambe et al[7] in Maiduguri [7] and Chukwu et al[35] in Enugu but slightly higher than Adegoke et al in Ife [23] and Akodu et al [36] in Lagos. In all these reports, [7, 23, 35, 36] at least three-quarter of the patients were diagnosed at age 5 and below. 6.8% of our patients were diagnosed at adolescents' age; this is far higher than the 2.1%, 2.2% and 1.9% from Maiduguri,[7] Enugu [35] and Lagos [36] respectively. The absence of routine neonatal screening for

SCD in the country is a major factor in the delay in diagnosis. [34, 35 36] Neonatal Screening has impacted significantly on SCD outcomes in the USA and Canada. [37, 38] High maternal educations, upper socioeconomic status, diagnosis of SCD in an elder sibling have been associated with early diagnosis in Nigeria. [35,36,39]

More children were being increasingly diagnosed with SCD since the inception of the health facility. There is paucity of data in Nigeria and indeed the African continent about trends in SCD diagnosis. Sanganuwan [28] in Sokoto didn't show trend in diagnosis over a 14 year study of SCD.

Makani et al[40] in Tanzania demonstrated more children being diagnosed with SCD over a 10 year period in a supported project in Muhimbili national hospital. While there is dearth of cohort studies amongst CLSCD in Nigeria, increasing health promotion and education, prenatal and genetic counseling, establishment of SCD clinics have impacted on health seeking behaviour. [35,36]

In our study, consanguinity was established in about a quarter of the parents of CLSCD. To the best of our knowledge this is the first report of consanguinity in CLSCD in the country. This occurred predominantly in parents with Muslim background. Consanguinity occurred more in Fulani ethnic group but was not statistically significant. There is paucity of reports on consanguinity amongst parents of CLSCD in Nigeria. Populations with high consanguinity marriages rates have significantly higher incidence of inherited blood disorders such as sickle cell anemia. [41,42]Reports [43,44] from the Middle East demonstrated the association of SCA with parental consanguinity.

While Muslims predominated in our study, on the other hand Christians constituted more than ninety percent of CLSCD in Ekiti South West Nigeria [45]. Nigerians of Fulani and Hausa ethnic background are largely of the Islamic Faith. [46]

More than half of CLSCD are of Fulani and Hausa ethnic background. While this may be related to population size of these ethnic groups in Gombe metropolis, it is probable consanguinity was underreported. Efforts of Saudi Arabia's government to provide genetic counselling services and implementing of mandatory premarital screening program have virtually eliminated SCD.[44]

History of siblings death in index cases was reported in about one fifth of families in our study. Sibling deaths from SCD were reported as early as 1917. [47] Saidi H et al [48] in Tanzania reported a family history of SCD death and its relation to stroke in SCD. However while this study did not seek to establish age at death, cause and place of death of a sibling there is paucity of reports of sibling death in Nigeria with the highest burden of the disease in Africa. It is probable these deaths may have been outside the health facility and of children less than 5 years, however this requires further studies.

A third of CLSCD were the 5th and beyond in order of birth. First, second and third orders of birth of CLSCD were almost equal in proportion. To the best our knowledge this is the first report of birth order of 5 and beyond among CLSCD of in Nigeria. Akodu et al[36,52] in two separate studies in Lagos had reported birth order in CLSCD but were limited to greater or equal to two. As an autosomal recessive disorder each birth from heterozygous parents carries equal risk of inheritance. [1,2]

About half of CLSCD belonged to the low socioeconomic class. This is similar to reports by Adegoke in Ife,[29] Ezenwosu [49] and Uchendu [50] both in Enugu but in contrast to findings of Akodu [51] and Animasahun in Lagos [16] and Rabiu in Kano [52] where majority of CLSCD were from the upper socioeconomic stratum. While several factors could account for this difference a systematic review [53] of published studies in past 20 years showed that children and adolescents with SCA predominantly belong to families from the less favored socioeconomic classes. Socioeconomic status is a factor that directly affects treatment and access to health care. [16,35,36]

Our study findings have both clinical and public health implications. Genetic counseling and legislation for mandatory premarital screening for SCD should be top priorities on the agenda of health. In addition neonatal screening and longitudinal cohort studies are required in order to enhance early diagnosis and deepen the epidemiologic context of this disease.

Acknowledgment

We wish to acknowledge Hajiya Fatima Y Aliyu and Hajiya Zainab Danmalam of the data unit of paediatrics department for extracting the data and all the staff of the Sickle Cell Centre for commitment to caring for these children.

Conflict of interest

Dr Elon Isaac is editor-in- chief for Gombe Medical journal. To ensure that any possible conflict of interest relevant to the journal has been addressed, this article was reviewed according to best practice guidelines of international editorial organizations.

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