



Otological findings among Nigerian children with sickle cell anaemia

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Summary

Background/Aim: Various degrees of hearing loss have been associated with sickle cell anaemia, especially of the sensorineural type (SNHL). However, there is little information on hearing pattern among sickle cell children in Nigeria. This study is to determine the prevalence of sensorineural hearing loss (SNHL) among children with sickle cell anaemia (SCA).

Patients and methods: Eighty (80) stable children aged 4–15 with Hbss attending the pediatric sickle cell clinic and also 60 control patients with HbAA, matched for age, sex at the pediatric general medical clinic of the University of Ilorin teaching hospital, Ilorin, Nigeria, all had prospective study of their pure tone audiological assessment (PTA) and tympanometric evaluations done over a year period.

Results: Their age range was 4–15 years with a mean of 9.4 for the Hbss and 9.7 for the control group. The male/female ratio was 1.3:1 and 1.5:1 for SCA and control subjects respectively.

25 subjects (50 ears) had abnormal audiograms among the SCA subjects and OME was the cause in 22 subjects and only three (3) had mild SNHL which was bilateral. However, in the control group 15 had abnormal audiograms and all were due to OME and none had SNHL. OME was bilateral in 19 subjects with SCA, two on the left and only one on the right. In the control group, 11 of the OME was bilateral and only four were on the left side. The prevalence of SNHL was 3.8% and OME was 27.5%.

Conclusion: We have found a prevalence rate for SNHL of 3.8% for 80 subjects with HbSS, and all cases have been a mild bilateral high frequency SNHL. Our findings

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suggested that SNHL is uncommon in early childhood, specifically during the years of language acquisition and early schooling. This could mean an age dependant prevalence rate of SNHL among SCA patients. However, no difference in the incidence of OME among both groups which can lead to educational difficulties from the resultant speech and language defects.

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1. Introduction

Sickle cell anaemia (SCA) was first described by Herrick in 1910 [1]. It is an inherited autosomal recessive disorder. The carrier state to have sickle cell trait (SCT) [2]. The prevalence of SCA is high among Africans and their descendants, Arabs, central and southern America and people of the Mediterranean area [1].

In African-Americans, the prevalence of SCT is 7–9%, and that of SCA is approximately 0.25%. It is a chronic hemolytic anaemia with vaso-occlusive episodes resulting in a variety of clinical features typical of SCA [1].

SCA is due to a single-point mutation in the 6th position of beta chain. The amino acid glutamic is replaced by valine, leading to changes in the physiochemical property of the red blood cells resulting in microvascular occlusion [1]. Data in Nigeria shows that 2 out of 100 young babies have sickle cell disease [3], an unpublished report by Akinyanju in Nigeria estimated that about 40 million Nigerians are carrying the sickle cell trait (SCT). The most significant clinical manifestations of SCA are those associated with CNS involvement [4].

The CNS is quite sensitive to even temporary hypoxia, for this reason, incidence of peripheral and central auditory dysfunction is higher among SCA than the normal population [4].

SNHL is one of the several complications associated with SCA [5–8] and it has been found to occur in 8% of SCA children in Nigeria, 12% in USA, 22% in Jamaica, 36.5% in Kenya and 60% in Ghana [3].

Morgenstein and Menace [9] in 1967 stated that compression of the auditory canal, by the expanded bone marrow of the petrous temporal bone and ischaemic damage to the hair cells of organ of corti of the cochlea due to microthrombotic occlusions of the cochlear venous system could lead to sensorineural hearing loss.

This study is to determine whether there is a higher prevalence of hearing disorders in children with SCA compared to the control with HbAA matched for age and sex attending the pediatric outpatient sickle cell clinic and the general pediatric clinic of the University of Ilorin Teaching Hospital, Ilorin, Nigeria.

2. Materials & methods

This prospective study was conducted over a 1 year period at the University of Ilorin teaching Hospital (U.I.T.H.), Ilorin, Nigeria between August 2005 and July 2006. Children with HbSS who are crisis free attending the pediatric outpatient clinic in steady state and controls with HbAA attending the general pediatric outpatient clinic were randomly selected and enlisted in the study. The control group was matched for age and sex and was documented to have normal Hb electrophoresis with HbAA pattern and no other haematological diseases. Ethical clearance was given by the hospital authority and individual consents secured from the parent/guardian of each child after explaining the purpose of the study.

Detailed history, otological and audiological examinations were carried out on both the randomly selected subjects and the controls and findings which included demographic data, clinical course of SCA were entered into a structured questionnaire administered by the investigators. The diagnosis of SCA (HbSS) was based on the clinical evaluation and hemoglobin electrophoresis.

Excluded were children with other diseases like mumps, meningitis, chronic suppurative otitis (CSOM), measles, drug ototoxicity or previous history of hearing loss, children with current sickle cell crises and those less than 4 years of age who are known to give inconsistent responses to audiometric testing. Otological and audiological examinations were conducted by the ENT consultants and senior registrars in addition to the evaluation by the consultant pediatrician and the senior registrars. Ears were examined and state of the tympanic membrane noted and tuning fork tests done with 512 Hz vibrating tuning forks (Rhine & Weber's tests) to detect possible hearing losses.

Evaluation consisted of pure tone audiometry, tympanometry and speech audiometry. The pure tone audiometer used was calibrated according to the American National Standard Institute, 1969 standard. Hearing levels were assessed separately in each ear in a sound proof booth at frequencies of 250, 500, 1000, 2000, 4000 and 8000 Hz. Measurements were made on a model dampex AS 67 diag-

nostic audiometer calibrated into ISI standard and expressed as decibels and impedance audiometer AT 235 DK 5610 Assens used for tympanometry.

A modified Jerger's classification of tympanograms was used as written below:

Type A—normal peak at 0	Type B—flat curve with no obvious peak (associated with OME)
Type AS—shallow peak	Type C—negative peak (associated with eustachian tube dysfunction)
Type Ad	Peak exceeds full scale on the ordinate

Audiological ambient noise level outside the booth was <30 dB measured by a sound meter. Pure tone thresholds of 25 dB or more in one or more test frequencies from 250 to 8000 Hz was considered a hearing loss for the purpose of this study. Similar evaluations were done for 60 age/sex matched children with HBAA. The PTA thresholds were characterized as conductive (CHL), sensorineural (SNHL) hearing losses. Hearing loss was categorized into mild, moderate, severe and profound and hearing thresholds for each frequency was obtained by simple averaging. Data were analyzed by EPI INFO 2005-3.3.2 Version comparison of variables done by student's t test and all analysis were evaluated for a level of significance of $p > 0.05$.

3. Results

Eighty (80) children homozygous HbSS (SCA) and 60 children without SCA (HbAA) participated in this study. From Table 1, the age range was 4–15 years with a mean age of 9.4 years (S.D. 3.5) for the HbSS and the mean age for the control was 9.7 (S.D. 3.4). The peak age group was 4–7 years with 55 children (68.7%), followed by 8–11 years with 18 children (22.50%) and lastly children 12–15 years of age being only 8.75%. The sex distribution among HbSS, showed 45 males (56.25%) and 35 females (43.75%) with a male/female ratio was 1.3:1.0 and the control group

showed a male/female ratio of 1.4:1.0, the age/sex distribution showed matching in both groups.

Parental occupations showed that more than half were civil servants 45 (56.25%), traders were 20 (25%) and others were 15 (18.75%). The parental educational background showed 42 had tertiary education (52.5%), 20 (2.5%) with primary education and 18 (22.5%) with secondary education. The social class classification based on socio-economic reasons and occupations, showed 48 (60%) were in the middle class, 26 (32.5%) in the lower class and only 6 (2.5%) in the upper class.

The school performances were average in 41 children (51.25%), good in 33 (41.25%), 5 children were exceptional (6.25%) and only a child had poor performances (1.25%). This educational/social class classifications were probably responsible for awareness on the nature of SCA, hence the importance of regular clinic visits and balanced diet.

The pure tone thresholds from Table 2, was normal bilaterally in 55 children (68.75%), however, there was evidence of conductive hearing loss (CHL) bilaterally in 19 children (23.75%) with a pure tone average of 30–40 dB; while 2 had left CHL (2.5%) with only a child (1.25%) with a right CHL. Bilateral sensorineural hearing loss (SNHL) with a pure tone average of 30–40 dB was found in only 3 children less than 7 years of age (3.8%) as found in Table 3. For the control group, 15 had evidence of bilateral CHL with a pure tone average of 30–35 dB in all the test frequencies and none had SNHL (Table 4).

Table 1 Age distribution of children with SCA and the control

Age group (years)	Frequency		Percentage (%)	
	HbSS	HbAA (control)	HbSS	HbAA
4–7	55	41	68.75	68.4
8–11	18	14	22.50	23.3
12–15	7	5	8.75	8.3
Total	80	60	100.00	100.00

Table 2 Age distribution/hearing loss among SCA/control children

Age group (years)	Frequency CHL		SNHL HbSS only
	HbSS	HbAA	
4–7	15 (60%)	10 (66.7%)	3 (12%)
8–11	5 (20%)	4 (26.7%)	—
12–15	2 (8%)	1 (6.6%)	—
Total	22	15	3

Table 3 Pure tone audiometric pattern among children with SCA and control

Hearing pattern	HbSS	HbAA
Normal bilateral	55	45
CHL (bilateral) ^a	19	11
CHL (left) ^a	2	4
CHL (right) ^a	1	—
SNHL ^b	3	—
Total	80	60

^a CHL: conductive hearing loss (mild to moderate degree).

^b SNHL: sensorineural hearing loss.

Table 4 Frequency distribution of hearing loss over 25 dB in both ears of HbSS and control groups

Frequency (Hz)	SS		Control	
	Right ear	Left ear	Right ear	Left ear
Conductive hearing loss (CHL)				
250	4	4	1	1
500	—	2	2	2
1 kHz	1	—	1	1
4 kHz	4	4	6	6
6 kHz	5	5	4	4
8 kHz	6	6	1	1
Total	Bil 19, RT 7, LT 1		15	15
Sensorineural hearing loss (SNHL) at 8 kHz				
	3	3	Nil response	

NB: Bil, bilateral; RT, right; LT, left.

The tympanometry was normal in 58 children (72.50%) and abnormal in 22 children (27.50%) with type B tympanograms in 19 children (23.75%) (16 was bilateral (20.0%); 2 on the left side (2.50%) and 1 on the right side (1.25%)) with the remaining 3 children with type As tympanogram (3.75%). All the 15 children with CHL in the control group had OME, all with type B tympanograms. Both the HbSS and HbAA children with OME had medical treatment with only two needing ventilation tube insertions and none needed hearing aid devices as there was no educational difficulties.

4. Discussion

Sickle cell anaemia (SCA) is a chronic hemolytic disorder, manifested by a variety of clinical and hematological features and complications, one of these is sensorineural hearing loss (SNHL) [4]. Med-line search variously reported increasing presence of SNHL among individuals with SCA compared to the general population, this could be due to impaired blood supply to the cochlea and the supporting cells along the peripheral and central auditory pathways [3]. The organ of hearing is not immuned from the hemolytic anaemia and blood vessel obstruction which are characteristics of homozygous sickle cell disease [6]. There is a consensus in the various audiological studies, that patients with SCA have a much higher incidence of SNHL than the general population [7]. Various series reported auditory impairment associated with sickle cell anaemia is due to the sickling and slugging of the cochlea (Diggs, 1956; Morganstein and Menace, 1969; Odetoyinbo and Adekile, 1987; Todd et al., 1973) [3].

Ischemia of the stria vascularis, with secondary organ of corti hypoxia as a direct result of sickle cell crisis, poses significant risk to the auditory system [3].

Large series report a prevalence of hearing loss of 11–41% in sickle disease patients [10]. Studies done in Nigeria among children by Odetoyinbo and Adekile [5] reported 21.4%, Mgbor and Emodi [3] reported 13.4% and Ogisi and Okafor [11] found the prevalence of SNHL among SCA children to be 8%, in contrast to studies from other parts of Africa, in Kenya and Ghana, with a reported higher prevalence of 36.5% and 60% respectively among SCA children [3].

Several reasons of age, locality and type of hemoglobin gene of the patient have been adduced for the varying prevalence rates of SNHL worldwide [3]. The prevalence rate of 3.8% is much lower than previous series from Nigeria, this could be due to the fact that the HbSS children in this series were crisis free and are seen regularly in the clinic. Also more than half of the parents were literate civil servants in the middle socio-economic class who value the importance of regular follow-up clinics and balanced diet for these children. Furthermore, a recent study by Aderibigbe et al. [8] at our centre reported a prevalence rate of 4.3% SNHL among adults with SCA, this was attributable to advances in medicine resulting in better care of SCA and general improvement in well-being of individuals with HbSS in Nigeria. Also in a study by Piltcher et al. in Brazil, reported a rising life expectancy among the populace due to earlier diagnosis of SCA coupled with better management of the condition [1].

Previous studies have noted that SNHL becomes more severe with increasing age and increasing number of sickle cell crisis of patients [6,7], but in this study, we did not find an increase in the frequency of hearing loss with increasing age as children below 8 years of age were mostly affected with those older than 12 years being only a tenth, Freidman et al. [12] could not confirm this statement in their study as well.

The sensorineural hearing loss in this series among the HbSS group was of mild type (30–40 dB) and was absent among the controls.

The pure tone hearing thresholds showed evidence of mild conductive hearing loss in 22 patients in all frequencies tested in the HbSS group. Conductive hearing loss (CHL) and SNHL were differentiated in this study as in MacDonald et al. [2], and nearly a third of the HbSS children with abnormal audiograms had CHL and OME was the etiology in over four fifth of them. This is similar to a study in the USA among cohort of children with SCA, middle

ear effusion was the etiology in 19 out of the 84 children screened and only 3 had mild SNHL [2]. OME is thought to be uncommon in Africans [13], as there have been only few reports on the prevalence of OME in Nigerian children. From a relatively low prevalence of 4.9% in Okeowo [14] and 8.9% in Ogisi series [15] to 23.8% in Akinlade et al. [16].

The findings in this study among the subjects and the control are similar to Akinlade et al. [16]. Most of the children were less than 7 years of age.

Literature on tympanometric findings among children with SCA is scanty. This finding shows OME is of the same frequency in both children with HbAA and HbSS.

In conclusion, we have found a prevalence rate of mild SNHL of 3.8%. Our findings suggested that SNHL is uncommon in early childhood specifically during the years of language acquisition and early schooling. This could mean an age dependant prevalence rate of SNHL among SCA patients. However, no difference in incidence of OME among children with SCA and those without. Regular audiometry should be done to monitor hearing thresholds in these patients to detect early hearing losses to avoid associated speech and language defects resulting in possible educational difficulties.

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