Hearing Thresholds in Sickle Cell Anemia Patients: Emerging New Trends?

Ademola Aderibigbe, MBBS, FWACP, FMCP; Foluwasayo E. Ologe, MBBS, FWACS, FMCORL; and Benjamin A. Oyejola, PhD
Ilorin, Nigeria

Purpose of study: Advances in medicine resulting in better understanding of sickle cell disease and general improvement of the well-being of the sufferers even in the developing countries have positively affected the dreadful outlook of this disease with resultant increase in the population of sickle cell disease patients reaching adulthood, and less severe complications. We therefore set out to evaluate the presence and severity of sensorineural hearing loss in sickle cell anemia (SCA) patients in the light of the overall improvement in the morbidity and mortality.

Methods: A prospective case control study of SCA patients attending our adult SCA clinic and control subjects from homozygous hemoglobin AA patients attending the staff clinic of the hospital for routine medical tests. Tympanometry and diagnostic audiometry were performed on each patient.

Main Findings: Forty-six SCA patients (21 males, 45.7%) aged 16–48 years with a mean age of 22.9 years ± 6.45 and 42 controls (24 males, 57.1%) aged 15–39 years with a mean age of 23.7 years ± 5.69 were included in this study. The average hearing thresholds of SCA patients were consistently higher than controls in all frequencies tested in both right and left ears. Of the 92 ears of SCA patients tested, 95.7% exhibited hearing thresholds within normal limits, and 4.3% had mild hearing loss. The controls had thresholds within normal limits.

Conclusion: The incidence of significant sensorineural hearing loss in SCA seems to have reduced in line with the general improvement and survival of SCA patients. The hearing loss is worse in the right ear and has a female preponderance. We hope that more aggressive primary and secondary prevention and adequate treatment of sickle cell crisis would reduce if not eliminate the hearing loss found in SCA.

Key words: hearing thresholds ■ sickle cell anemia ■ new trends

 From Department of Medicine, University of Ilorin Teaching Hospital (Aderibigbe, consultant physician); and the Department of Otolaryngology, College of Medicine (Ologe, senior lecturer/consultant otolaryngologist) and Department of Statistics (Oyejola, professor and head), University of Ilorin, Ilorin, Nigeria. Send correspondence and reprint requests for *J Natl Med Assoc.* 2005;97:1135–1142 to: Dr. F.E. Ologe, PO Box 6641, Ilorin, Nigeria; e-mail: foluologe@yahoo.com

INTRODUCTION

Sickle cell anemia (SCA) patients are known to have worse morbidity and mortality compared to their age- and sex-matched controls in the general population. An exception is the increased resistance of patients with sickle cell trait against malaria. Also, SCA patients have been observed to have a lower arterial blood pressure than controls.[1]

Previous reports have shown a much higher prevalence of sensorineural hearing loss in adult SCA patients in the malarial-endemic tropical regions[2-4] than the developed countries.[5] Reasons given for this are related to the severity of the course of the disease due to the specific hematological profile, certain geographical factors and the level of medical care available.[2-7]

Advances in medicine resulting in better understanding of sickle cell disease and general improvement of the well-being of the sufferers even in developing countries have positively affected the dreadful outlook of this disease with resultant increase in the population of sickle cell disease patients reaching adulthood, and less severe complications.[4,8,9] We therefore set out to evaluate the presence and severity of sensorineural hearing loss in SCA patients in the light of the overall improvement in the morbidity and mortality of SCA patients.[4,8,9]

METHODS

This prospective study was carried out among homozygous sickle cell anemia patients attending the medical outpatient (MOP) clinic of the University of Ilorin Teaching Hospital, Ilorin, Nigeria regularly for at least the preceding year.

Patients were recruited from the MOP clinic after careful explanation of the research procedure and the time constraints involved. Their informed consent was sought and obtained. They were referred to the audiology clinic of the hospital where they were evaluated using a structured questionnaire. History of previous ear disease; ear or head trauma and/or surgery; exposure to excessive noise; ototoxic drugs,

and diseases, such as meningitis, mumps and measles, were grounds for exclusion from the study.[10,11] For the purpose of this study, apart from a detailed history of present and past occupations and noise-related hobbies, we used the questions:

A) Are you exposed to noise at work or during leisure time?
B) If yes, do you consider this noise level high enough to constitute a risk of noise induced hearing loss?

Answer to question B was recorded as a measure of the awareness of exposure to harmful noise.[12,13] Those answering yes to question B were excluded from the study.

After otoscopy, only patients with clear external auditory canals and intact and shiny tympanic membranes proceeded further in the study. However, patients with wax impaction had gentle syringing of the ear, and further audiological measurements were delayed for at least one week.

Tympanometry was done with an impedance tympanometer AT 235 (Interacoustics, Denmark) following standard procedures.[10-11,14] Only patients with normal tympanograms, i.e., type-A, proceeded to audiometry. A diagnostic audiometry was performed on each patient by a clinical audiologist in a double-walled, soundproof cabin, using a duly calibrated (Testo 815) diagnostic audiometer (Danplex AS 67 by GN Otometrics A/S, Denmark) with well-fitting TDH 35 earphones employing standard procedures.[10,11,14] Measurements of pure tone thresholds by air conduction were obtained at 0.125-, 0.25-, 0.5-, 1-, 2-, 4- and 8 kHz in each ear. Also, bone conduction measurements were done with an appropriate bone vibrator placed on the respective mastoid bone at 0.25-, 0.5-, 1-, 2- and 4 kHz in each ear.

Pure tone averages (calculated for 0.5-, 1-, 2- and 4 kHz) were classified into one of the following categories: Within normal limits (-10 dB–25 dBHL), mild hearing loss (26–40 dBHL), moderate hearing loss (41–55 dBHL), moderately severe hearing loss (56 dB–70 dB), severe hearing loss (71–90 dBHL) and profound hearing loss (>91 dBHL).[2-4,15] Patients with air-

Table 1. Age, sex and occupational distribution

Parameter	Frequency (%) Subjects	Controls	Statistics
Age (Years)			χ^2=2.66;df=2; P value=0.26
15–25	39 (84.8)	30 (71.4)	
26–35	5 (10.9)	10 (23.8)	
>35	2 (4.3)	2 (4.8)	
Total	46 (100)	42 (100)	
Sex			χ^2=1.16; df=1;P value=0.28
Male	21 (45.7)	24 (57.1)	
Female	25 (54.3)	18 (42.9)	
Total	46 (100)	42 (100)	
Occupation			
Student	32 (69.6)	39 (92.9)	
Fashion designer	5 (10.8)	—	
Trader	5 (10.8)	1 (2.4)	
Civil servant	1 (2.2)	2 (4.7)	
Clergyman	1 (2.2)	—	
Photographer	1 (2.2)	—	
Barber	1 (2.2)	—	
Total	46 (100)	42 (100)	

Table 2. Average hearing thresholds of subjects and controls (using t test)

Frequency kHz	Average Hearing Threshold (dB ± SD)		Statistics	
	Subjects (N=46)	Controls (N=42)	t Value‡	P Value
R 0.125	22.3 ± 11.4	15.1 ± 11.5	2.933	0.004*
R 0.25	24.5 ± 10.8	14.5 ± 11.6	4.172	0.0001*
R 0.5	21.7 ± 10.0	13.1 ± 11.2	3.832	0.0001*
R 1.0	16.5 ± 16.0	10.4 ± 9.7	2.165	0.03*
R 2.0	14.6 ± 16.5	10.2 ± 10.4	1.455	0.1
R 4.0	13.9 ± 17.5	10.7 ± 10.5	1.027	0.3
R 8.0	19.7 ± 17.9	13.0 ± 11.6	2.062	0.04*
RR	16.7 ± 13.0	11.1 ± 9.1	2.320	0.023*
L 0.125	21.2 ± 12.2	15.0 ± 9.8	2.614	0.01*
L 0.25	20.2 ± 9.9	14.9 ± 8.4	2.719	0.008*
L 0.5	19.7 ± 11.1	12.1 ± 7.7	3.652	0.0001*
L 1.0	12.2 ± 10.3	9.4 ± 6.3	1.511	0.1
L 2.0	9.0 ± 10.0	7.7 ± 7.0	0.690	0.5
L 4.0	10.4 ± 12.9	9.2 ± 8.2	0.546	0.6
L 8.0	14.4 ± 13.9	10.7 ± 9.9	1.398	0.2
LL	12.8 ± 9.4	9.6 ± 5.8	1.900	0.06

R: right ear; L: left ear; RR: average of R 0.5, R1.0, R 2.0, R 4.0; LL: average of L 0.5, L1.0, L 2.0, L 4.0; * statistically significant difference; ‡ df: 86

bone gap of ≥15 dB, regarded as having conductive hearing loss,[10,11] were excluded from the study.

Age- and sex-matched controls with hemoglobin AA were taken from individuals who came for routine medical tests in the same hospital. They went through the same exclusion criteria, examination and audiological procedures as the SCA patients.

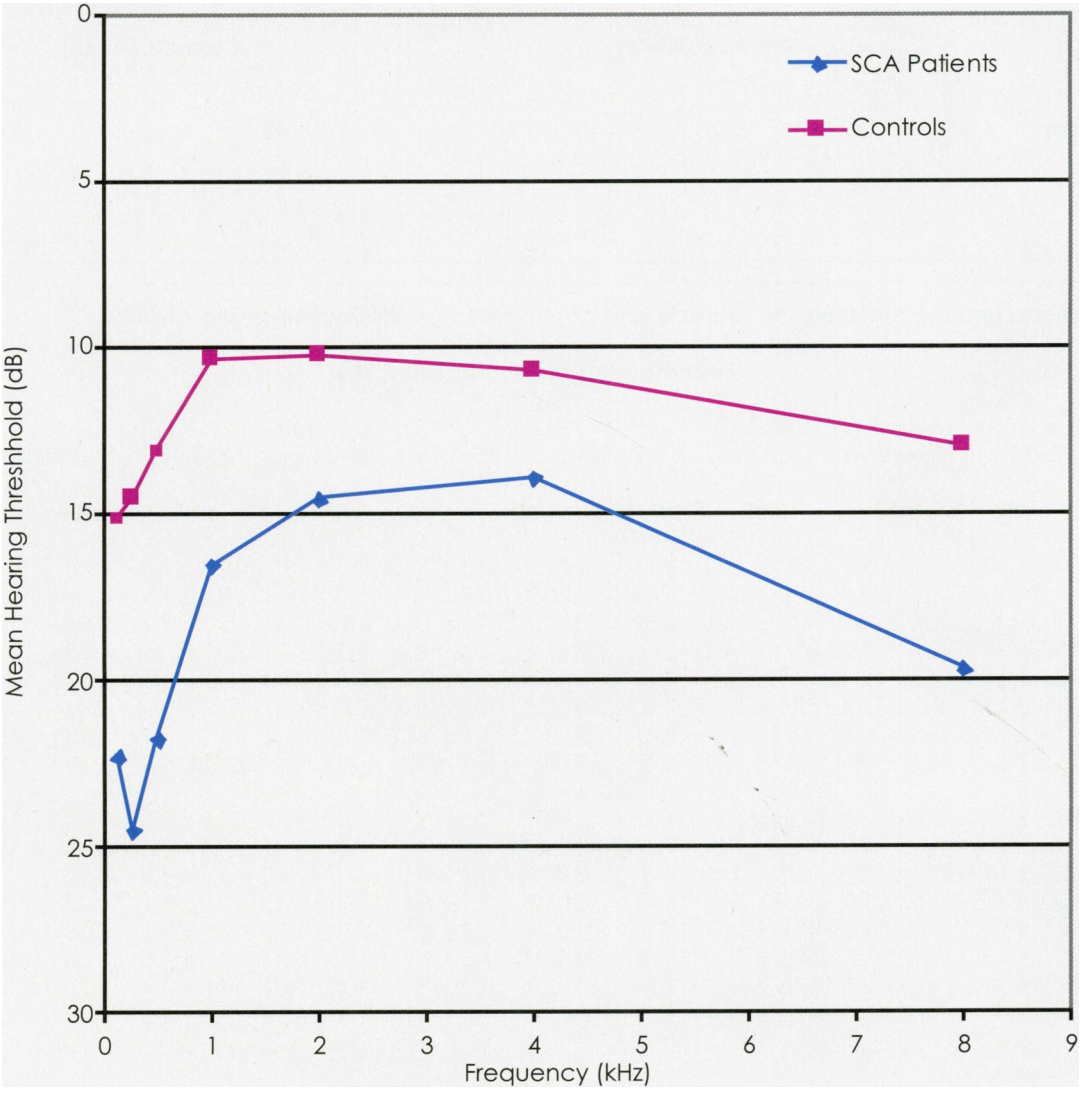
Since air- and bone conduction thresholds were similar in included subjects, only air conduction audiograms were analyzed. Results were analyzed by comparison of group means ± standard deviation using two-tailed t tests. Analysis of variance (ANOVA) was used to assess the impact of age, gender and other variables of interest on hearing threshold. Differences were considered statistically significant only if p value <0.05.

RESULTS

Forty-six SCA patients (21 males, 45.7%) aged 16–48 years with a mean age of 22.9 years ± 6.45 and 42 controls (24 males, 57.1%) aged 15–39 years with a mean age of 23.7 years ± 5.69 were included in this study. The Chi-squared test shows that age and sex distribution of the subjects are similar to those of controls, i.e., no statistically significant differences in age and sex distribution (Table 1). This is an indication of matching, although not in its simplest applied form (i.e., paired matching), where a one-for-one pairing occurs between the population of interest and the control group. One-hundred percent and 94% of the controls and SCA patients, respectively, had at least secondary education; 93% and 70%, respectively, were still in school. Only two of the SCA patients gave a positive history of hearing loss.

The average hearing thresholds of SCA patients

Figure 1. Mean hearing threshhold (right ear) in patients and controls



were consistently higher than controls in all frequencies tested in both right and left ears. The t test identified that the differences in the threshold between SCA patients and controls were statistically significant in both low and high frequencies in the right ear and in the lower frequencies in the left ear. Also, there is an indication of differences in the hearing threshold in the right and left ears. Using a t test, the hearing threshold in the right ear tended to be worse than in the left ear, the difference being statistically significant at 0.25 kHz (t=2.613; df=45; p value= 0.01), 2.0 kHz (t=2.292; df=45; p value=0.03), and 8.0 kHz (t=2.061; df=45; p value=0.045) in SCA patients. This analysis in controls was not statistically significant (Table 2, Figures 1 and 2).

Of the 92 ears of SCA patients tested, 95.7% exhibited hearing thresholds within normal limits; and 4.3% had mild hearing loss. Of the 84 ears of control subjects, all exhibited hearing thresholds within normal limits (Table 3).

In comparing the subjects in age groups of 10-year intervals, ANOVA showed that there was a statistically significant difference in the hearing threshold with advancing age at all frequencies tested except 1.0 kHz in the right ear. Among the control group, although the hearing threshold generally increased with advancing age, ANOVA did not show a statistically significant difference in the thresholds. (Table 4).

Female SCA patients consistently had worse hearing thresholds than their male counterparts in all frequencies tested in both right and left ears. However, using t test, this difference was only statistically significant in lower frequencies (0.125 kHz–0.5 kHz) in both ears and at 8.0 kHz in the left ear. Also, female controls generally had worse hearing thresholds than their male counterparts in all the frequencies tested

Table 3. Category of hearing threshold

Hearing Threshold	Frequency (%)					
	Subjects (N=46)			Controls (N=42)		
	R	L	Total	R	L	Total
Within normal	43	45	88 (95.7)	42	42	84 (100)
Mild hearing loss	3	1	4 (4.3)	—	—	
Total		92 (100)			84 (100)	

R: right ear; L: left ear

Table 4. Mean hearing thresholds in subjects and controls by age distribution (using ANOVA)

Frequency kHz	Mean Hearing Thresholds (dB ± SD)									
	Subjects					Controls				
	15–25 yrs	26–35 yrs	>35 yrs	Statistics		15–25 yrs	26–35 yrs	>35 yrs	Statistics	
	n=39	n=5	n=2	F value*	P value	n=30	n=10	n=2	F value‡	P Value
R 0.125	20.4 ± 10.4	32.0 ± 9.1	35.0 ± 21.2	4.11	0.023*	16.3 ± 11.2	11.0 ± 13.1	17.5 ± 3.5	0.85	0.437
R 0.25	22.7 ± 9.3	30.0 ± 9.4	45.0 ± 21.2	5.88	0.006*	15.7 ± 11.4	10.0 ± 12.5	20.0 ± 7.1	1.14	0.330
R 0.5	19.7 ± 8.4	30.0 ± 10.6	40.0 ± 14.1	7.59	0.002*	13.8 ± 11.4	9.5 ± 11.2	20.0 ± 7.1	0.96	0.393
R 1.0	15.1 ± 14.6	18.0 ± 22.5	40.0 ± 14.1	2.48	0.096	10.2 ± 8.4	8.5 ± 9.7	22.5 ± 24.8	1.85	0.171
R 2.0	13.7 ± 15.8	9.0 ± 12.9	45.0 ± 7.1	4.27	0.020*	9.5 ± 9.3	10.5 ± 13.0	20.0 ± 14.1	0.96	0.394
R 4.0	12.4 ± 15.7	9.0 ± 13.4	55.0 ± 14.1	7.50	0.002*	10.5 ± 9.3	10.0 ± 13.9	17.5 ± 10.6	0.44	0.648
R 8.0	17.4 ± 15.2	18.0 ± 14.8	67.5 ± 3.5	10.67	0.001*	13.5 ± 11.2	11.0 ± 13.9	15.0 ± 7.1	0.20	0.821
RR	15.3 ± 11.5	16.5 ± 13.4	45.0 ± 12.4	6.17	0.004*	11.0 ± 8.1	9.6 ± 10.9	20.0 ± 14.1	1.10	0.344
L 0.125	20.3 ± 12.0	20.0 ± 7.9	42.5 ± 3.5	3.57	0.037*	15.8 ± 9.1	10.5 ± 10.9	25.0 ± 7.1	2.34	0.110
L 0.25	18.6 ± 8.8	24.0 ± 8.2	42.5 ± 3.5	7.77	0.001*	15.0 ± 7.8	14.0 ± 11.0	17.5 ± 3.5	0.15	0.861
L 0.5	18.3 ± 10.5	22.0 ± 10.4	40.0 ± 7.1	4.27	0.020*	12.7 ± 8.0	10.5 ± 8.0	12.5 ± 3.5	0.29	0.753
L 1.0	11.0 ± 9.5	13.0 ± 5.7	32.5 ± 17.7	4.92	0.012*	9.2 ± 6.4	10.0 ± 6.2	10.0 ± 7.1	0.07	0.930
L 2.0	7.2 ± 7.6	11.0 ± 4.2	40.0 ± 14.1	18.10	0.001*	7.8 ± 6.5	7.0 ± 8.9	10.0 ± 7.1	0.16	0.856
L 4.0	8.3 ± 7.6	8.0 ± 9.1	57.5 ± 17.7	35.45	0.001*	9.3 ± 7.9	8.0 ± 10.1	12.5 ± 3.5	0.26	0.769
L 8.0	11.9 ± 10.6	16.0 ± 6.5	57.5 ± 17.7	17.91	0.001*	10.2 ± 8.7	11.5 ± 14.0	15.0 ± 7.1	0.26	0.777
LL	11.2 ± 7.1	13.5 ± 2.0	42.5 ± 14.1	18.56	0.001*	9.8 ± 5.7	8.8 ± 6.8	11.3 ± 3.5	0.16	0.851

R: Right ear; L: left ear; RR: average of R 0.5, R1.0, R 2.0, R 4.0; LL: average of L 0.5, L1.0, L 2.0, L 4.0; * statistically significant difference; * df: 2 & 43; ‡ df: 2 & 39

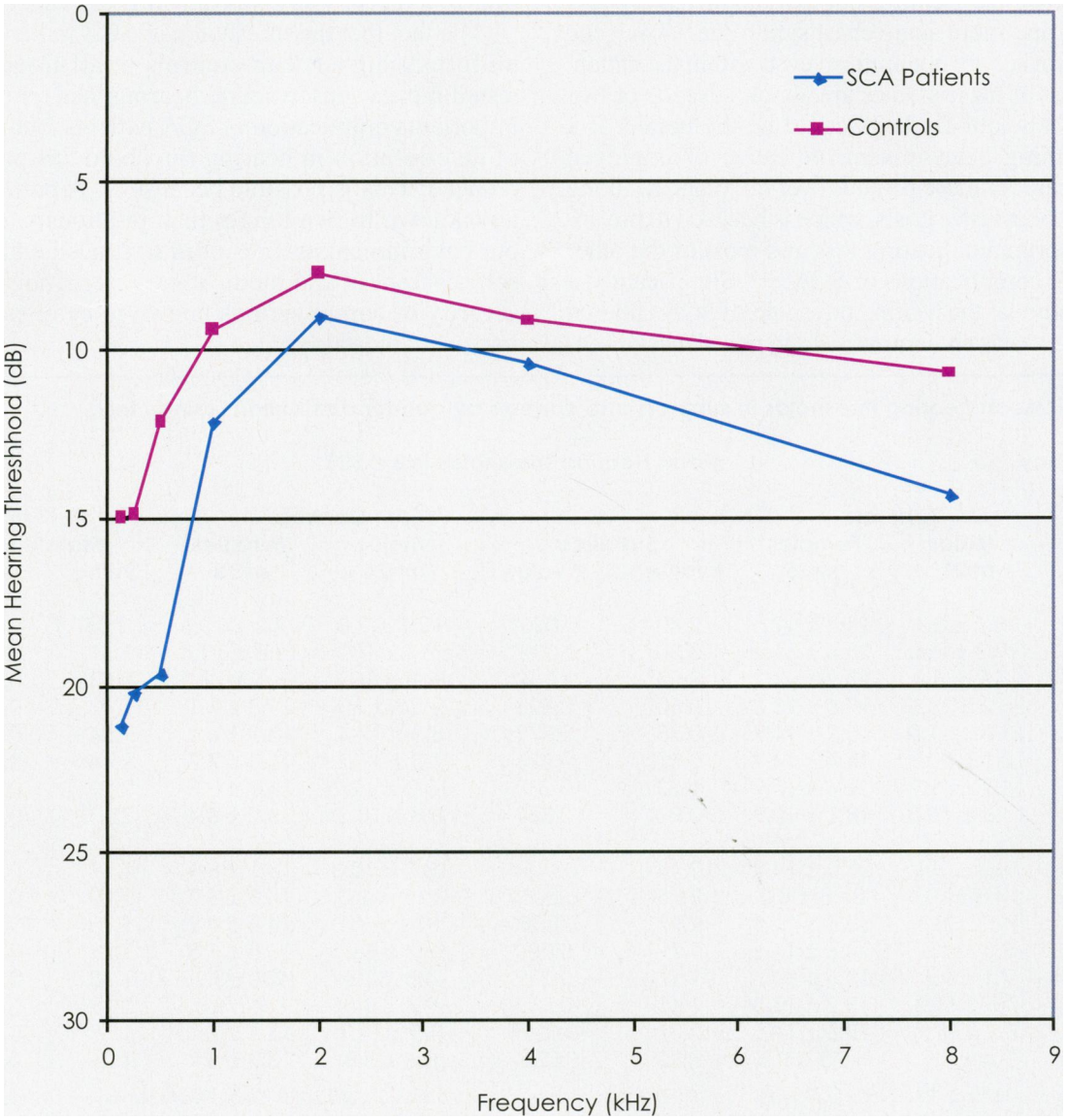
except at 4.0 kHz in the left ear. Using t test, differences were statistically significant only at 0.125 kHz and 4.0 kHz in the right ear and at 0.25 kHz in the left ear. The average hearing threshold was worse for females in both ears for both SCA patients and controls. However, t test showed a statistically significant difference in this threshold in the left ear for SCA patients and in the right ear for controls (Table 5).

Table 6 is a summary of ANOVA for age, sex and group differences in hearing thresholds in SCA patients and controls. It shows that the hearing threshold increases significantly with advancing age at all frequencies except 0.125 kHz in both right and left ears in the SCA patients and controls. Also, females (both SCA patients and controls) had significantly worse hearing thresholds at 0.125–0.5 kHz and 8.0 kHz in the right ear and in all except 4.0 kHz in the left ear. Group differences are as stated earlier.

DISCUSSION

Most studies have shown significantly increased hearing threshold in SCA patients,[2-7,16,17] but there is no audiometric pattern that could be regarded as pathognomonic.[18] While occasional reports of conductive hearing loss in SCA patients have been published,[19] most workers have observed sensorineural hearing loss affecting all frequencies.[2-5,7,17,20] Significantly worse high-frequency thresholds have been reported by some,[3,6,21] and both lower and higher frequencies by others.[2] In the present study, the SCA patients had

Figure 2. Mean hearing threshhold (left ear) in patients and controls



consistently worse hearing thresholds at all frequencies tested, although the audiometric patterns were similar to that of the control subjects. Also, the differences in their hearing thresholds were statistically significant at both high and low frequencies in the right ear and at lower frequencies in the left.

In our study, which is the very first of its kind in Ilorin (a melting point in terms of vegetation and population representation in Nigeria, corresponding to the northern parts of the south, and the southern part of the north), only 4.3% of the ears had mild sensorineural hearing loss. This proportion is quite lowm compared to the 30–66% reported for adult SCA patients with similar age range in our environment.[2-4,21] This difference could be due to differing specific hematological profiles of SCA in various locations. In a previous study comparing the clinical profiles of SCA patients in Lagos (southwestern Nigeria) and Benin (mid-western Nigeria), the Benin group had significantly less dactylitis and more acute chest syndrome.[22] Thus, the clinical profiles are not uniform even within the nation. Also, most of these studies are about a decade or two old.[2,3,21] Thus, our findings could be the herald of a heartwarming development indicative of improved medical care for these patients over the years, resulting in less vasoocclusive crisis, which is believed to precipitate sensorineural hearing loss and most of the other associated complications of SCA.[6,21,23] Significantly, a recent study[16] of the hearing threshold of SCA children in Nigeria shows an improvement in prevalence of sensorineural hearing loss over a similar study done almost two decades ago.[6] Experience in our medical center shows that with improved formal education and health awareness, SCA patients tend to be more regular with clinic appointments, comply better with their routine medication and report earlier to the hospital on noticing symptoms of possible complications (having been adequately tutored). Overall, these make for better health.

Our findings could also be a reflection of improved preventive measures against other causes of sensorineural hearing loss in our environment, which were additive to the specific pathogenesis of sensorineural hearing loss in SCA patients. The latter view could be valid because the hearing thresholds among the controls in the present study were much better than in earlier reports.[2-7,16,17,21] Also, the prevalence of sensorineural hearing loss among controls of a recent study was only 6.2%.[16]

The fact that the thresholds of SCA patients were uniformly higher than controls at all frequencies tested means sensorineural hearing loss remains an important complication of SCA patients, but the rate of deterioration in hearing threshold has probably reduced. This is possible because SCA patients are now known to live longer than previously, even in our environment as a result of improved education, self-awareness and medical care, resulting in less severity of complications known to be responsible for earlier mortality.[4,8,9]

Table 5. Mean hearing thresholds in subjects and controls by gender distribution (using t test)

Frequency kHz	Mean Hearing Thresholds (dB ± SD)							
	Subjects				Controls			
	Males	Females	Statistics		Males	Females	Statistics	
n	n=21	n=25	t value*	P value	n=24	n=18	t value‡	P value
R 0.125	18.1 ± 8.4	25.8 ± 12.5	2.40	0.020*	12.1 ± 9.8	19.2 ± 12.6	2.05	0.047*
R 0.25	19.3 ± 7.6	28.8 ± 11.2	3.30	0.002*	12.1 ± 10.4	17.8 ± 12.5	1.61	0.116
R 0.5	17.9 ± 6.8	25.0 ± 11.1	2.57	0.014*	10.8 ± 10.1	16.1 ± 12.2	1.53	0.133
R 1.0	15.2 ± 18.5	17.6 ± 13.8	0.50	0.623	8.5 ± 11.0	12.8 ± 7.1	1.43	0.162
R 2.0	13.8 ± 21.0	15.2 ± 11.9	0.28	0.779	8.1 ± 12.4	13.1 ± 6.2	1.54	0.131
R 4.0	12.1 ± 20.9	15.4 ± 14.4	0.62	0.536	7.5 ± 11.2	15.0 ± 7.7	2.44	0.019*
R 8.0	16.2 ± 19.9	22.6 ± 15.9	1.22	0.230	10.0 ± 12.7	16.9 ± 8.8	2.00	0.053
RR	14.8 ± 15.3	18.1 ± 10.9	0.92	0.362	8.8 ± 10.2	14.2 ± 6.4	2.13	0.040*
L 0.125	16.7 ± 7.1	25.0 ± 14.2	2.44	0.019*	12.7 ± 10.3	18.1 ± 8.4	1.79	0.080
L 0.25	14.8 ± 8.3	24.8 ± 8.8	3.95	0.001*	12.7 ± 9.0	17.8 ± 6.7	2.10	0.042*
L 0.5	14.8 ± 8.7	23.8 ± 11.4	2.97	0.005*	10.4 ± 7.1	14.4 ± 8.2	1.70	0.096
L 1.0	9.3 ± 7.6	14.6 ± 11.6	1.79	0.080	8.3 ± 6.0	10.8 ± 6.5	1.29	0.205
L 2.0	7.1 ± 9.6	10.6 ± 10.3	1.17	0.249	6.5 ± 7.6	9.4 ± 5.9	1.38	0.174
L 4.0	7.1 ± 11.0	13.2 ± 13.8	1.62	0.112	9.4 ± 8.5	8.9 ± 8.0	0.19	0.852
L 8.0	9.3 ± 11.7	18.6 ± 14.5	2.37	0.022*	8.5 ± 11.1	13.6 ± 7.4	1.68	0.102
LL	9.4 ± 7.9	15.5 ± 9.8	2.22	0.031*	8.6 ± 5.8	10.9 ± 5.7	1.26	0.215

R: Right ear; L: left ear; RR: average of R 0.5, R1.0, R 2.0, R 4.0; LL: average of L 0.5, L1.0, L 2.0, L 4.0; * statistically significant difference; * df: 44; ‡ df: 40

The occurrence of sensorineural hearing loss significantly increased with advancing age in this study. This is in agreement with most studies[4,17,20] but at variance with a suggestion that the crucial period in the development of sensorineural hearing loss in sickle cell disease may be intrauterine or during the first few years of life.[5] The clinical significance of the foregoing is that our SCA patients may eventually develop more severe degrees of sensorineural hearing loss but hopefully at a much older age. Hence, there is a need for periodic evaluation of the hearing status of SCA patients as part of their general care. Also, adequate counseling is necessary to avoid exposure to other factors, particularly excessive noise, that may contribute to or aid progression of sensorineural hearing loss.

We also observed that the hearing thresholds of female SCA patients and controls were significantly worse relative to their male counterparts at all except 4.0 kHz in the left ear and at 0.125–0.5kHz, as well as 8.0 kHz in the right. Hitherto the literature has been essentially silent on gender differences in the hearing threshold of SCA patients except for a few report of equal sex prevalence.[2] In the general population, current studies are inconclusive regarding specific patterns of gender differences in sensorineural hearing loss.[24-26] However, results show that hearing sensitivity declines more than twice as fast in men as in women at most ages and frequencies and that >1 kHz males show greater average loss than females, but <1 kHz females show greater average loss than males.[24,26] On the whole, the male sex may be associated with increased incidence of hearing loss after adjusting for age.[25] One of the major reasons adduced is related to more noise exposure, with the attending noise-induced hearing loss in males. However, in the present study, excessive noise exposure was one of the exclusion criteria. Where facilities for an objective assessment are not available, as in many developing countries, appropriate questions addressing noise exposure (like those used in this study) have been found to be useful and reliable alternative means for screening subjects exposed to high noise levels (>85 dB).[12,13] Specifically, in SCA patients, it may not be inconceivable to suggest that menstrual blood loss, known to be partly responsible for lower packed-cell volume in females, may put additional hemodynamic stress on female SCA patients and probably predispose them to worse cochlea damage during vasoocclusive crisis.

The present study has its limitations. The design cannot adequately answer the question of new trends in hearing threshold measurement relative to improved medical care of SCA patients in developing countries. A comparison within SCA patients based upon 10-year age intervals would probably be more illustrative of this hypothesis, i.e., if the overall treatment of SCA patients is better, then one would expect to see fewer differences in hearing loss as an SCA patient moved from decade to decade. Even this proposition would have to bear in mind the confounding factor of the influence of age on hearing thresholds, particularly as this study shows that age has a greater impact on the hearing thresholds of SCA patients than on controls. Furthermore, the etiopathogenesis of hearing loss in SCA patients is not exactly fully settled. Also, our controls were not technically matched, although they were not significantly different from the experimental group. Moreover, 33% of the matching cells contain a count of <5 against the assumption of the Chi-squared analysis test, which requires that no more than 20% of cells have an expected count of <5 to support validity.[27,28] Comparing the severity of SCA disease (detailed analysis of indices, such as serial packed-cell volume, frequency and severity of vasoocclusive crisis, as well as other complications) with hearing threshold is the subject of an ongoing study (consequent on our present observations) aimed at further assessing the current status of the well-being of SCA patients in our environment.

Table 6. Analysis of variance between subjects and controls

Frequency (kHz)	Age (Years)		Sex		Group	
	F Value‡	P Value	F Value‡	P Value	F Value‡	P Value
R 0.125	2.253	0.1	10.127	0.002*	8.205	0.005*
R 0.25	4.503	0.04*	12.362	0.001*	18.204	0.0001*
R 0.5	8.147	0.005*	9.326	0.003*	16.628	0.0001*
R 1.0	7.179	0.009*	2.037	0.2	5.140	0.03*
R 2.0	7.577	0.007*	1.580	0.2	2.685	0.1
R 4.0	9.109	0.003*	3.528	0.06	1.665	0.2
R 8.0	12.854	0.001*	5.915	0.02*	5.343	0.02*
L 0.125	3.498	0.07	9.241	0.003*	7.119	0.009*
L 0.25	10.112	0.002*	20.305	0.0001*	9.348	0.003*
L 0.5	5.921	0.02*	12.592	0.001*	14.620	0.0001*
L 1.0	6.696	0.01*	5.985	0.02*	2.394	0.1
L 2.0	17.825	0.0001*	5.079	0.03*	0.833	0.4
L 4.0	14.087	0.0001*	2.370	0.1	0.773	0.4
L 8.0	21.688	0.0001*	11.869	0.001*	3.186	0.08

R: right ear; L: left ear; * statistically significant difference; ‡ df: 1

In conclusion, the hearing threshold of SCA patients is worse than age- and sex-matched controls. However, there appears to be a decline in the occurrence of socially significant sensorineural hearing loss in line with the general improvement and survival of SCA patients. It was also observed that hearing loss worsened with advancing age and female gender. We do hope this work will generate sufficient interest to encourage a larger population based study on this subject and hopefully be an encouragement to continue to strive for better health for SCA patients.

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DEDICATION

This work is affectionately dedicated to the blessed memory of Grace Oluwafunmilayo Oyejola, who passed on to glory on May 24, 2005.

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