Original article

EVALUATION OF HEARING THRESHOLDS OF CHILDREN AND ADOLESCENTS WITH SICKLE CELL DISEASE AGED 5 – 16 YEARS IN AHMADU BELLO UNIVERSITY TEACHING HOSPITAL, ZARIA, NIGERIA

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Abstract

Context: Sickle cell disease (SCD) causes abnormal hemoglobin, leading to complications such as vaso-occlusive crises, chronic anemia, and organ damage. One significant but underreported complication is sensorineural hearing loss (SNHL), which often starts with gradually elevated hearing thresholds and can lead to permanent hearing loss. This study aimed to compare the hearing thresholds of children and adolescents with SCD to those with normal hemoglobin (HbAA).

Method and Materials: This hospital-based, cross-sectional case-control study evaluated 125 children and adolescents (ages 5 to 16 yrs) with sickle cell disease (subjects) in a steady state, compared to 125 healthy controls with haemoglobin AA genotype. Subjects were recruited from the haematology outpatient clinic, while controls were selected from the general outpatient clinic at Ahmadu Bello University Teaching Hospital (ABUTH).

Results: The mean hearing thresholds in both ears for SCD subjects (20.0±2.5 dB; t=12.918; p<0.001) were significantly higher than those for controls with the HbAA genotype (14.2±3.4 dB; t=12.918; p<0.001). Adolescents with SCD exhibited elevated mean hearing thresholds at both low and high frequencies, with an increase in hearing thresholds as they aged. There was no significant difference in mean hearing thresholds between genders (Males: 21.1 dB; Females: 21.2 dB).

Conclusion: The study concluded that children and adolescents with sickle cell disease (SCD) have significantly higher hearing thresholds than their peers with normal HbAA genotype. This hearing loss

increases with age and affects both genders equally. These findings underscore the importance of regular hearing evaluations and targeted care for individuals with SCD.

Keywords: Hearing threshold, Sensorineural hearing loss, Sickle cell disease, Steady stat

Introduction

Sickle cell disease (SCD) refers to a collection of genetic blood disorders characterized by the presence of a haemoglobin variant called sickle cell haemoglobin (HbS). It is the most common autosomal recessively inherited genetic disease and the most debilitating haematological genetic disorder affecting the black race today.[1]. The global burden of sickle cell disease (SCD) remains significant and is increasing. The total number of people living with SCD worldwide has increased by 41.4%, reaching approximately 7.74 million in 2021. Nigeria plays a significant role in the global burden of SCD. As one of the countries with the highest incidence rates, it is part of a group that accounts for nearly half of the global SCD incidence at birth. Efforts to manage and mitigate the impact of SCD (of which hearing loss is one but underreported) in these regions are crucial, given the high mortality rates among children under five and the significant health disparities that exist.[1-3]

Hearing thresholds refer to the quietest sound that can be heard by an individual, measured in decibels (dB) and is used to determine the degree of hearing loss. The degree of hearing loss a person has can be characterized by how loud a sound has to for the person to be able to hear it. Normal hearing threshold in children and adolescents is defined as 15 decibels (dB) or better (less). In children and adolescents with sickle cell disease (SCD), studies have consistently shown that these thresholds are often elevated compared to their peers with normal hemoglobin (HbAA). [4-12]

Hearing loss in sickle cell disease (SCD) results from several interrelated mechanisms influenced by the disease's

pathology.[13-17] One primary factor is vaso-occlusion and ischemia, where the sickling of red blood cells leads to blockages in small blood vessels, including those supplying the cochlea. This causes ischemia and subsequent cochlear damage, resulting in sensorineural hearing loss (SNHL).[18-20] Chronic anemia, another key aspect of SCD, exacerbates this issue by reducing the oxygen-carrying capacity of the blood. The inner ear is particularly sensitive to oxygen deprivation, and prolonged hypoxia can damage cochlear hair cells, further contributing to hearing loss.[21, 22] Compounding this problem, children with SCD are prone to recurrent infections due to impaired spleen function. Frequent ear infections (otitis media) can initially cause conductive hearing loss, and severe or repeated infections can lead to permanent sensorineural damage.[23-27] Additionally, opioids such as hydrocodone, oxycodone, morphine, and codeine are frequently used to manage moderate to severe vaso-occlusive crises (VOCs) in SCD. There have been reports of rapid-progression hearing loss associated with hydrocodone abuse, and a few cases have identified hearing loss as an adverse effect of opioid use.[28-30] These findings highlight the need for cautious use of these common drugs in SCD to avoid exacerbating hearing loss. Moreover, chronic blood transfusions, a common treatment for SCD, can lead to iron overload. Excess iron deposits in various organs, including the cochlea, cause oxidative stress and cellular damage that impair hearing.[31-33] Finally, genetic factors may play a role in the variability of hearing loss among SCD patients, as certain genetic variants may increase susceptibility to hearing damage, explaining why some patients experience more severe hearing loss than others.[20,34-39] Understanding these interconnected mechanisms underscores the need for comprehensive care and regular auditory evaluations in individuals with SCD to detect and manage hearing loss early, thereby improving their quality of life.[40]

Overall, Children and adolescents with sickle cell disease (SCD) tend to have higher hearing thresholds and a greater prevalence of sensorineural hearing loss compared to their peers with normal hemoglobin.[41-43] The Northwest region of Nigeria significantly contributes to the national SCD burden, yet there are few studies on the hearing thresholds of affected individuals in this area. This study aims to evaluate the hearing thresholds of children and adolescents with SCD in comparison to their peers with normal hemoglobin patterns.

Methods

Study Design and Setting

A hospital-based, cross-sectional case-control study was conducted at Ahmadu Bello University Teaching Hospital (ABUTH) in Zaria, North-West Nigeria. ABUTH's Sickle Cell Clinic, which serves Zaria and nearby areas, sees 60 – 70 children with SCD weekly.

Study Population and Source

The study included children and adolescents aged 5-16 years with sickle cell disease (Subjects) from the Sickle Cell Clinic and those with normal hemoglobin (controls) from the General Outpatient Department at Ahmadu Bello University Teaching Hospital (ABUTH), Zaria. Participants were those who attended these clinics during the study period.

***Inclusion criteria for subjects and controls*:** Subjects: Patients with documented hemoglobin genotypes SS, SC, or SS+F and regular clinic follow-up; Controls: Individuals with an HbAA genotype. Consent or assent was obtained before recruitment.

Exclusion criteria for subjects and controls: Children with SCD in acute crisis or with other hemoglobinopathies. Those with a family history of hearing loss, ear surgery, head trauma, radiotherapy, stroke, chronic blood transfusions, excessive noise exposure, diabetes,

HIV, TB, measles, mumps, meningitis, or ototoxic drug intake.

Sample Size Estimation

The required sample size was calculated using the single population proportion formula,[44] assuming a 95% confidence interval (CI), a 5% margin of error, and an 8% proportion of children and adolescents with elevated hearing thresholds – sensorineural hearing loss (based on a study by Ogisi *et al.*[15] in Southeastern Nigeria). This initial calculation yielded a sample size of 113. After accounting for a 10% non-response rate, the final sample size was determined to be 125. Hence, the sample size calculated was 125 subjects with Sickle cell disease and 125 age- and -sex matched controls.

Sampling Technique

Subjects meeting the inclusion criteria were randomly selected using a random number table. An average of ten participants (subjects and controls) were recruited weekly over six months (June–December, 2021). Recruited subjects underwent Pure Tone Audiometry (PTA) and Tympanometry on the same day. Age and sex-matched controls from the outpatient departments had their haemoglobin genotype determined before the audiologic tests.

Data Collection

A pretested, structured interviewer-administered questionnaire was used to collect data from subjects and their caregivers, covering bio-data, socio-demographic characteristics, and hearing loss symptoms. The diagnosis of SCD (HbSS, HbSC, or HbSS+F) for subjects was verified from case notes, while controls (HbAA) had their haemoglobin genotype confirmed by electrophoresis during the study.

Audiologic Evaluation: An otolaryngologist conducted a thorough physical examination of both study subjects and controls using an otoscope to rule out any middle ear infections, impacted earwax, or ear perforations. Only individuals with clear ear canals and intact

eardrums were included in the study. If any patients had impacted earwax, it was removed, and their auditory testing was postponed for one week before proceeding. Middle ear function in all participants, including controls, was evaluated using tympanometry by two clinical audiologists, each possessing over 10 years of experience—one from the National Ear Center and the other from Ahmadu Bello University Teaching Hospital. The assessment was conducted with a TYMP 87 Clinical Middle Ear Analyzer, manufactured by GN Otometrics in Copenhagen, Denmark. Only those with normal Type-A tympanograms and an acoustic reflex, indicating normal middle ear function, were allowed to continue with audiometric testing. Diagnostic audiometry was carried out by the clinical audiologists using a calibrated audiometer (MADSEN ITERA, also by GN Otometrics, Copenhagen, Denmark), along with properly fitting TDH 35 earphones. Testing took place in a double-walled, soundproof cabin to ensure accuracy. The audiometer was pre-calibrated for the frequencies of 125 Hz, 250 Hz, 500 Hz, 1 KHz, 2 KHz, 4 KHz, 6 KHz, and 8 KHz, and air conduction measurements of pure tone thresholds were recorded for each ear across these frequencies. Hearing loss was classified based on WHO criteria, with the average hearing threshold between 500 Hz and 4000 Hz used to categorize loss as mild, moderate, moderate-severe, severe, or profound..[45]

Data Analysis

Data entry and analysis were performed using IBM SPSS statistical software version 25 (IBM Corporation, Armonk, NY, USA). Student t-test or one-way analysis of variance (ANOVA) was used to determine the statistical significance of mean values of hearing thresholds. Differences were considered statistically significant only if p value <0.05.

Ethical Clearance

Institutional approval with ethical clearance (ABUTH/HREC/K6/2021) was obtained

from the Health Research Ethics Committee of Ahmadu Bello University Teaching Hospital, Zaria, Nigeria. Written informed consents and assent were obtained from the participants and parents/caregivers of the participants.

Results

Socio-demographic characteristics of participants

Of the 125 subjects aged 5-16 years, 68 (54.4%) were males and 57 (45.6%) were females, with a male-to-female ratio of 1.2:1 and a mean age of 10.17 ± 3.35 years. The Chi-squared test indicated no significant differences in age and sex distribution between subjects and controls (X^2 = 0.75; P = 0.69), showing effective matching. Among the subjects, 85 (68%) had homozygous HbSS, while 6 (4.8%) had HbSC and HbSS + F accounted for 34 (27.2%). All controls were HbAA. The majority of study participants were of Hausa ethnicity (84.8%), followed by Yoruba (7.2%), Igbo (1%), and others (7.2%). Among the subjects with sickle cell anaemia, two-thirds (83.3%) were from a low socioeconomic class. Additionally, 38.4% of the subjects had poor follow-up clinic attendance, and 43.2% demonstrated poor adherence to routine medication regimens.

Mean Middle Ear Volumes in Subjects and Controls

The mean middle ear volumes for both the right and left ears among subjects (Right Ear: 0.74 ± 0.10 mL; Left Ear: 0.73 ± 0.12 mL) and controls (Right Ear: 0.72 ± 0.09 mL; Left Ear: 0.71 ± 0.07 mL) show no significant statistical differences. For the right ear, the t-test value is t=1.660, p=0.09; for the left ear, the t-test value is t=1.610, p=0.10, with a degree of freedom (df) of 248. These results indicate that there is no significant difference in middle ear volume between the subjects and controls.

Mean Hearing Thresholds in Subjects and Controls

Subjects with sickle cell disease (SCD) consistently had higher mean hearing thresholds than controls at all tested

frequencies in both ears (Figure 1). SCD subjects perceived spoken words at high normal thresholds (20.0 ± 2.5 dB in the right ear; 20.2 ± 2.5 dB in the left ear) compared to controls, who had normal hearing thresholds (14.2 ± 3.6 dB in both ears) across all frequencies and age groups.

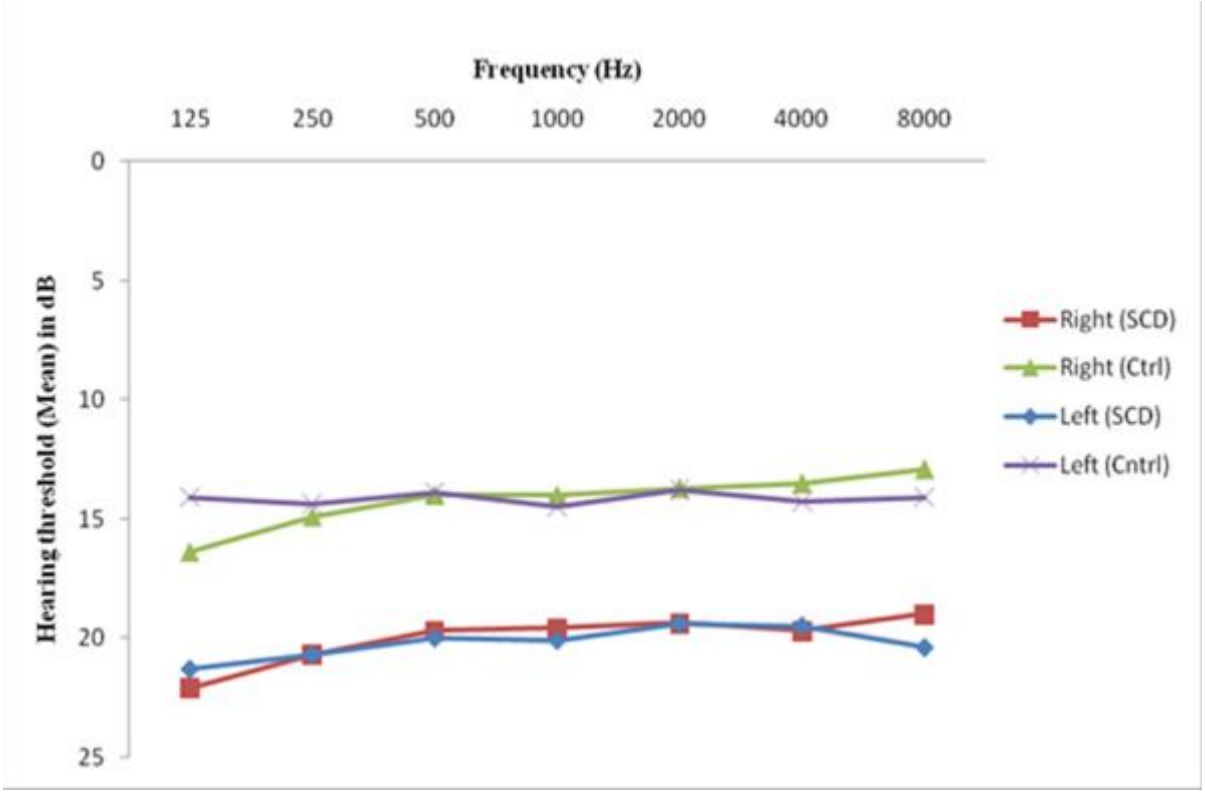

Figure 1: Mean PTA Hearing Thresholds of SCD Subjects and Control HbAA

Table 1: Comparison of Mean Hearing threshold in Subjects and Controls by Age distribution

	Mean hearing thresholds in subjects and controls by age distribution (using ANOVA)									
	Mean Hearing Thresholds (dB ± SD)					Mean Hearing Thresholds (dB ± SD)				
	Sickle cell disease (n=125)					**Controls (Hb AA) n=125**				
Frequency	**(5 – 8) yrs**	**(>8 – 12)yrs**	**(>12-16)yrs**	**Statistics**		**(5 – 8) yrs**	**(>8– 12)yrs**	**(>12-16)yrs**	**Statistics**	
(Hz)	n=45	n=47	n=33	**F value[ɣ]**	**P value**	n=45	n=47	n=33	**F value[ɣ]**	**P value**
R 125	21.5 ± 3.4	22.5 ± 4.7	24.9 ± 4.7	2.76	0.067	16.3 ± 2.9	16.6 ± 3.7	16.0 ± 3.1	0.46	0.633
R 250	20.6 ± 2.9	20.7 ± 3.6	24.3±10.4	4.49	0.013*	15.3 ± 4.0	15.0 ± 4.0	14.0 ± 4.0	0.86	0.427
R 500	19.4 ± 2.7	21.0 ± 4.0	22.6±11.4	2.19	0.116	14.2 ± 3.3	14.2 ± 3.2	13.2 ± 3.5	1.00	0.369
R1000	19.8 ± 2.7	20.6 ± 5.1	23.0±12.3	1.94	0.148	13.6 ± 3.2	14.6 ± 3.6	13.6 ± 3.7	1.20	0.305
R2000	19.8 ± 2.7	20.8 ± 7.1	23.9±13.4	2.52	0.085	13.4 ± 3.5	13.4 ± 3.9	14.3 ± 3.8	0.62	0.542
R4000	20.3 ± 3.7	21.0 ± 7.1	24.9±15.9	2.51	0.085	12.9 ± 3.6	13.3 ± 3.7	14.8 ± 4.2	2.89	0.059

R8000	20.0 ± 5.2	22.1 ± 9.7	27.4±15.4	4.88	0.009*	12.6 ± 3.6	13.1 ± 3.9	13.0 ± 4.6	0.15	0.858
L 125	21.5 ± 3.6	22.6 ± 3.5	23.1 ± 3.2	2.27	0.108	13.9 ± 4.1	13.9 ± 4.1	14.7 ± 4.8	0.50	0.610
L 250	20.6 ± 3.8	21.6 ± 3.6	21.7 ± 3.2	1.26	0.288	13.5 ± 3.0	14.8 ± 4.0	15.1 ± 4.3	2.11	0.125
L 500	20.0 ± 2.9	20.8 ± 4.3	21.2 ± 3.5	1.17	0.315	14.0 ± 3.0	14.0 ± 3.4	13.7 ± 3.3	0,09	0.909
L1000	20.0 ± 2.7	20.8 ± 5.0	21.6 ± 4.4	1.71	0.186	14.7 ± 3.0	14.7 ± 3.5	13.8 ± 4.0	0.97	0.382
L2000	19.9 ± 2.3	19.9 ± 7.2	21.8 ± 4.3	1.77	0.175	14.1 ± 2.9	13.7 ± 3.3	13.8 ± 3.2	0.42	0.660
L4000	20.2 ± 3.4	21.2 ± 7.8	22.6 ± 6.1	1.51	0.226	14.5 ± 3.6	14.4 ± 3.8	13.7 ± 3.7	0.55	0.579
L8000	20.1 ± 4.1	23.4 ± 8.9	25.3 ± 9.2	3.63	0.030*	13.3 ± 3.6	14.8 ± 3.8	14.3 ± 3.8	1.83	0.165

**Statistically significant difference; R = Right ear; L = Left ear; dB = decibel of hearing; SD = Standard deviation; ᵾdf = 3 and 122*

Table 2 : Mean Hearing threshold in subjects and controls by Gender distribution

	Mean Hearing Threshold (dB±SD) Using Student t-test							
	Sickle Cell Disease		**Statistics**		**Controls (HbAA)**		**Statistics**	
Freq	**Male**	**Female**			**Male**	**Female**		
(Hz)	n = 68	n = 57	t value	pvalue	n = 68	n = 57	t value	P value
R 125	22.7 ± 7.8	22.9 ± 4.2	- 0.216	0.829	16.6 ± 3.5	16.1 ± 2.9	0.932	0.353
R 250	21.7 ± 7.7	21.6 ± 3.6	0.130	0.897	14,2 ± 3.6	15.7 ± 3.8	- 2.342	0.021*
R 500	21.3 ± 8.4	20.1 ± 3.4	0.842	0.402	13.4 ± 3.5	14.6 ± 3.1	- 1.899	0.060
R1000	21.4 ± 8.8	20.4 ± 4.9	0.766	0.445	13.7 ± 3.6	14.1 ± 3.4	- 0.805	0.423
R2000	21.7 ± 9.9	20.7 ± 6,6	0.613	0.940	13.8 ± 4.0	13.6 ± 3.4	0.304	0.762
R4000	22.4 ± 11.6	21.0 ± 6.6	0.825	0.411	13.9 ± 3.9	13.0 ± 3.7	1.297	0.197
R8000	23.7 ± 12.2	21.7 ± 8.4	1.046	0.297	13.1 ± 4.0	12.7 ± 3.9	0.657	0.513
L 125	22.5 ± 3.5	22.1 ± 3.5	0.702	0.484	14.3 ± 4.3	13.9 ± 4.3	0.508	0.613
L 250	20.9 ± 3.2	21.7 ± 4.0	- 1.263	0.210	14.4 ± 4.0	14.5 ± 3.6	- 0.137	0.891
L 500	20.0 ± 3.1	21.4 ± 4.1	- 2.243	0.027*	13.8 ± 3.1	14.1 ± 3.3	- 0.454	0.651

L1000	20.6 ± 2.9	20.9 ± 5.0	- 0.520	0.604	14.6 ± 3.2	14.3 ± 3.8	0.527	0.599
L2000	20.3 ± 2.9	20.5 ± 7.0	- 0.262	0.794	13.4 ± 3.1	14.2 ± 3.3	- 1.348	0.180
L4000	21.6 ± 5.0	20.7 ± 7.1	0.408	0.455	13.8 ± 3.5	14.8 ± 3.9	- 1.457	0.148
L8000	23.9 ± 7.3	22.2 ± 8.3	1.245	0.215	14.0 ± 3.8	14.3 ± 3.9	- 0.337	0.737

**Statistically significant difference; degree of freedom= 123; dB = decibel of hearing; SD = Standard deviation; R = Right ear; L = Left ear; Freq = Frequency*

Mean Hearing Thresholds in Subjects and Controls by Age Distribution

Table 1 shows that in subjects with sickle cell disease (SCD), mean hearing levels increase with age at all frequencies in both ears. In contrast, children with normal hemoglobin (controls) consistently perceived spoken words at normal hearing levels (10-15 dB) in both ears across all age groups. Among controls, mean hearing levels did not significantly increase with age at any frequency.

Mean Hearing Thresholds in Subjects and Controls by Gender Distribution

The overall mean hearing thresholds for the male subjects with SCD were 21.1±9.5 dB Right ear; and 21.1±4.0 dB Left Ear while for the females, it was 21.2±5.4 dB Right Ear and 21.1±5.6 dB in Left Ear. For the control groups the total mean hearing thresholds for the male were 14.1±3.6 dB Right Ear; and 14.0±3.6 dB Left Ear while for the female it was 14.3±3.7 dB in either ear. The mean hearing thresholds by gender distribution showed no statistically significant difference among subjects with SCD and among the control group across the frequencies tested except for isolated R250Hz (t = - 2.342; P = 0.021 Right Ear) for control and L500HZ (t = - 2.243; p = 0.027) for the subjects (Table 2).

Discussion

All participants, including controls, demonstrated a 'Type A' tympanogram, characteristic of healthy ears, which indicates normal ear canal volume, an intact

tympanic membrane, properly functioning ossicles, and the presence of an acoustic reflex. This finding effectively rules out middle ear pathologies such as otitis media, perforations, or abnormal middle ear pressure. Consequently, the hearing threshold abnormalities observed in this study were confined to the inner ear structures, specifically indicating sensorineural hearing impairment. This impairment reflects the typical otologic manifestations seen in individuals with sickle cell disease, compared to those with normal hemoglobin (HbAA), underscoring the primary etiologic role of sickle cell disease in affecting auditory function.[19, 36, 42, 46, 47]

This study evaluated the hearing thresholds of children and adolescents with sickle cell disease (SCD) in comparison to those with normal hemoglobin (HbAA). The results revealed that the mean hearing thresholds for subjects with SCD were significantly higher than those of the control group, with average thresholds in both ears measuring 20.0 ± 2.5 dB for SCD subjects, while controls averaged 14.2 ± 3.4 dB. This finding indicates a notable elevation in hearing thresholds among children with SCD, which is corroborated by several studies.[17, 36, 42, 46] Yasir Nuhu Jibril *et al*.[7] in Nigeria assessed cochlear function in children with SCD and reported abnormal cochlear function, specifically elevated hearing thresholds compared to their healthy counterparts, supporting the current study's results. Similarly, Tantawya *et al*.[48] found that children and adolescents with SCD exhibited inner ear complications, including higher hearing thresholds, reinforcing the evidence of auditory dysfunction in this population. Rissatto-Lago *et al*.[8] conducted a systematic review and meta-analysis that highlighted dysfunction of the auditory system in individuals with SCD, further validating the observed elevated hearing thresholds. This comprehensive analysis indicates a broader trend of auditory impairment in SCD

patients across various studies. Additionally, Olajuyin *et al*.[10] documented significant otological burdens in Nigerian children with SCD, noting considerable hearing loss consistent with the findings of the current study, which underlines the widespread nature of this complication within the SCD population in the region. Furthermore, Stuart and Smith[49] examined the emergence and prevalence of hearing loss in children with homozygous SCD, discovering that these children are at a heightened risk of elevated hearing thresholds. The evidence from multiple studies confirms the increased vulnerability of the SCD population to hearing impairment, emphasizing the need for ongoing monitoring and intervention for auditory dysfunction in this group.

The elevated hearing thresholds in subjects with sickle cell disease (SCD) affect both ears equally, indicating a bilateral hearing impairment. This condition can be attributed to the systemic nature of SCD, where vaso-occlusive events, chronic anemia, and recurrent infections have a uniform impact on both ears. Rissatto-Lago *et al*.[8] highlighted in their systematic review and meta-analysis that auditory system dysfunction in SCD is typically bilateral, further reinforcing the observations of this study. Similarly, Tantawya *et al*.[48] found that inner ear complications in SCD patients are generally bilateral, which supports the systemic impact of the disease on hearing. Bois *et al*.[12] also reported bilateral sensorineural hearing loss in children with SCD, emphasizing the uniform nature of auditory impairment across this population. Furthermore, the study by Olajuyin *et al*.[10] indicated that Nigerian children with SCD commonly experience bilateral hearing loss due to the pervasive effects of the disease on the auditory system. Additionally, Yasir Nuhu Jibril *et al*.[7] confirmed that cochlear function is equally affected in both ears of SCD patients, aligning with the observed bilateral nature of hearing loss. Finally,

Stuart *et al*.[49] noted that hearing loss in children with SCD is typically bilateral, consistent with the overall pattern of systemic complications associated with the disease. However, it is important to acknowledge that some authors[16, 42, 50, 51] have reported unilateral elevated hearing thresholds in cohorts of children and adolescents with SCD, although this pattern is not typical given the mechanisms of hearing loss in this group.

The study revealed a significant influence of age on hearing thresholds, with older children with sickle cell disease (SCD) exhibiting elevated thresholds at both low and high frequencies. This trend aligns with the findings of Emilie Bois *et al*.[12] who observed progressive hearing loss in older children with SCD, suggesting that the cumulative effect of SCD-related complications exacerbates hearing impairment over time. Tantawya *et al*.[48] similarly found that inner ear complications, including elevated hearing thresholds, became more pronounced as children with SCD aged, indicating that prolonged exposure to the pathophysiological impacts of the disease worsens auditory function. Yasir Nuhu Jibril *et al*.[7] also highlighted the progressive nature of cochlear damage in SCD patients, noting that older children experienced more significant hearing loss compared to their younger counterparts. Rissatto-Lago *et al*.[8]confirmed through a systematic review and meta-analysis that the severity of auditory dysfunction in SCD patients correlates with age, further validating the observations of this study. Additionally, Olajuyin *et al*.[10] found that older Nigerian children with SCD exhibited more significant hearing loss, emphasizing age as a critical factor in the progression of auditory impairment. Heather K. Schopper *et al*.[52] supported these findings, noting higher hearing thresholds in older children with SCD in the United States, underscoring the pervasive nature of age-related hearing loss in SCD across different

regions. However, in contrast to these findings, Friedman *et al*.[53] (USA), Alabi *et al*.[42] (Nigeria), Ajulo *et al*.[54] (United Kingdom), and Al-Dabbous *et al*.[36] (Qatif, Saudi Arabia) did not observe an increased incidence of elevated hearing thresholds with advancing age among their studied subjects. This discrepancy suggests that age-related hearing loss might not be universal and indicates that other factors, such as genetic variability or differing healthcare standards, could influence hearing outcomes. This highlights the complexity of auditory impairment in SCD and underscores the need for further research to explore these discrepancies.

Gender did not significantly influence the mean hearing thresholds in this study, as both male and female subjects with sickle cell disease (SCD) exhibited similar hearing thresholds, which were higher than those of their HbAA counterparts. This lack of gender disparity is consistent with findings from Alabi *et al*.[42], who also reported no significant difference in hearing loss between male and female children with SCD. Similarly, Todd *et al*.[55] found that gender did not play a significant role in hearing threshold variations among children with SCD, and Tantawya *et al*.[48] corroborated these findings, indicating no notable gender differences in hearing loss within their cohort of SCD patients. Mgbor *et al*.[17] observed that the prevalence and severity of hearing loss were comparable between male and female children with SCD, further supporting the current study's conclusions. Olajuyin *et al*.[10] provided additional confirmation by reporting no gender-based differences in the auditory profiles of Nigerian children with SCD, and Alison *et al*.[46] found that both genders experienced similar degrees of hearing impairment, reinforcing the idea that gender does not significantly impact hearing thresholds in children with SCD. However, in contrast to these findings, later studies by Aderibigbe *et al*.[41], Onakoya *et al*.[56], and Al-Okbi *et al*.[43] described

poorer audiometric test results in female subjects compared to male subjects with SCD. Aderibigbe *et al*.[41] suggested that menstrual blood flow, which is partly responsible for lower packed-cell volume in females, may place additional hemodynamic stress on female patients with SCD, potentially predisposing them to worse cochlear damage during vaso-occlusive crises. Despite this, the proportion of adolescents in our study who had attained menarche was very small (0.2%), making it difficult to draw definitive conclusions about this hypothesis. We propose that these varying findings may be due to a complex interplay of factors influencing care, as well as differences in the quality and access to optimal care, which may have been difficult to fully assess across these studies.

Conclusion

This study demonstrates that children and adolescents with sickle cell disease (SCD) have significantly higher mean hearing thresholds compared to their peers with normal hemoglobin (HbAA), with hearing impairment affecting both ears equally. The findings suggest that the cumulative effects of SCD-related complications over time exacerbate hearing loss, particularly in older children. Additionally, the study highlights the socioeconomic disparities among SCD patients, with the majority coming from low socioeconomic backgrounds, coupled with poor follow-up attendance and medication non-adherence. These factors may further compound the health challenges faced by these individuals, emphasizing the need for a more comprehensive and accessible approach to SCD care. No significant gender differences were observed in hearing thresholds, reinforcing the systemic and pervasive nature of hearing loss in SCD patients across different demographic groups.

Limitation of the Study

The study had several limitations that should be acknowledged. Firstly, the hospital-based nature of the research may introduce sample bias, as participants were drawn from a specific clinical setting. Also, the study focuses on particular outcomes, such as hearing improvement, and does not explore quality-of-life measures or psychosocial impacts, it might provide an incomplete picture of the true burden and treatment efficacy for patients with SCD.

Recommendations

Given the findings, it is essential that routine audiometric evaluations are integrated into the standard care for children and adolescents with SCD, particularly those from lower socioeconomic backgrounds who may be more vulnerable to poor follow-up and medication non-compliance. Healthcare systems must develop targeted interventions to improve clinic attendance and adherence to medication regimens, such as community-based health outreach programs and caregiver education. Multidisciplinary care involving pediatricians, haematologists, audiologists, and social workers is crucial to addressing both the medical and socioeconomic challenges these patients face. In addition, healthcare providers should be trained to recognize the unique needs of SCD patients, ensuring that both auditory and general health outcomes are monitored closely. Policymakers should also prioritize accessibility to routine evaluations and medications, possibly through subsidies or national healthcare programs. Finally, further research is needed to explore the interaction between socioeconomic status, healthcare access, and the progression of hearing loss in SCD patients, as well as the potential role of genetic predispositions in determining the severity of auditory complications.

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Conflicts of interest

There are no conflicts of interest.

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