

Auditory Impairment in Children and Adults with Sickle Cell Disease and Sickle Cell Trait

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ABSTRACT

Background: Auditory impairment is one of the systemic consequences linked to sickle cell disease (SCD) and sickle cell trait (SCT). Nevertheless, little is known about the degree and nature of hearing loss in adults and children with SCD and SCT.

Aim: To evaluate the impact of hearing loss on children and adults with SCD and SCT, assess its severity, and identify potential risk factors.

Patients and Methods: This cross-sectional study was conducted at Al-Ahrar Teaching Hospital on 120 participants who were divided into four groups: 30 children with SCD, 30 adults with SCD, 30 children with SCT, and 30 adults with SCT. Comprehensive audiological assessments were performed to determine the presence and severity of hearing loss. Demographic data, including age and gender distribution, were also analyzed.

Results: The study revealed significant differences in hearing impairment between SCD and SCT groups. Children with SCD exhibited greater vulnerability to auditory dysfunction compared to their adult counterparts. The gender distribution showed a higher prevalence of hearing loss among females in the pediatric SCD group (56.7%), while adult SCD patients had a male predominance (66.7%). SCD was associated with more severe auditory and cognitive deficits than SCT.

Conclusion: SCD poses a significant risk for hearing loss, particularly in children, necessitating early screening and intervention to mitigate developmental and quality-of-life impairments. Further research is needed to identify the underlying pathophysiological mechanisms and optimize management strategies.

Keywords: Sickle Cell Disease, Sickle Cell Trait, Hearing Loss, Auditory Impairment, Cognitive Function.

INTRODUCTION

Black people frequently suffer from sickle cell disease (SCD) and sickle cell trait (SCT), which are genetically inherited abnormalities of the red blood cell. The global meta-estimate prevalence of sickle cell disease (SCD) was 1.12% in Africa, 0.04% in Europe, and 0.11% worldwide ⁽¹⁾. If left untreated, sickle cell disease (SCD) can lead to end-organ damage, vaso-occlusive crises, persistent hemolytic anemia, and an elevated risk of infections. One of the end organs that is at risk is the cochlea (organ of hearing). Wide differences in hearing impairment, particularly high-frequency sensorineural hearing loss, can be caused by a vaso-occlusive event that affects the cochlea's end artery supply ⁽²⁾.

Due to their heterozygous genetic origin, people with SCT are typically clinically stable; but, under severe situations like infections, dehydration, and exercise, they may experience a subclinical vaso-occlusive crisis. As a result, SCT may cause hearing loss, however there is little data on the consequences of this hemoglobinopathy and little research on those who have it ⁽³⁾.

Physical activities including playing, taking care of themselves, and doing housework may be restricted for

kids with SCD. Hospital stays and exacerbations can make it difficult for children to attend school and can impair their physical, mental, and cognitive abilities, especially when it comes to learning and attention ⁽⁴⁾.

The purpose of this study was to determine the degree of hearing loss, its effects on children and adults with SCD and SCT, and possible risk factors.

PATIENTS AND METHODS

This cross-sectional study aimed to determine how hearing loss affected adults and children with sickle cell trait (SCT) and sickle cell disease (SCD) and conducted at Al-Ahrar Teaching Hospital, General Organization for Teaching Hospitals and Institutes (GOTHI).

Inclusion Criteria

Adults (18-50 years) and children (ages 5–17) with hemoglobin electrophoresis-confirmed diagnosis of sickle cell disease (SCD) (HbSS, HbSC, HbS β^0 , or HbS β^+). Via hemoglobin electrophoresis, sickle cell trait (HbAS) was verified. No hospitalization or ongoing crises within the four weeks before enrollment.

Exclusion Criteria

1. A history of meningitis and head trauma, previous loud noise exposure and use of ototoxic medications during the previous six months.
2. A history of hearing loss unrelated to SCD or SCT.
3. Reliable audiometric testing is hindered by cognitive impairment.
4. Long-term illnesses unrelated to SCD that may impair hearing
5. Those who are in unstable or emergency situations were not allowed to participate.

All patients were subjected to full history taking, family history of hearing loss, health background including the genotype of SCD, the age at diagnosis, complications of disease, the frequency of crises of pain, hospitalizations, history of blood transfusions, present prescription drugs and the usage and duration of hydroxyurea.

Laboratory studies were done including complete blood count, electrophoresis of hemoglobin, ferritin in serum, tests for kidney and liver function and inflammatory indicators (ESR, CRP).

Equipments and materials:

- The two-channel audiometer (Madsen Itera) was used for pure-tone audiometry and calibrated according to the manufacturer's specifications. Immittance meter (MAICO model MI34) was used and calibrated according to the manufacturer's specifications.
- Auditory evoked potential acquisition system, ABR: Biologic navigator pro Natus to perform P300
- Locally made sound-treated room.

Audiological Assessment

- Otoscopy: Visual examination of the external auditory canal and tympanic membrane to rule out wax, infection, or structural abnormalities.
- Tympanometry: Conducted using a calibrated impedance audiometer to assess middle ear function and classify tympanometric curves (Types A, B, C).
- Pure-Tone Audiometry (PTA): Air and bone conduction thresholds were measured at frequencies of 250–8000 Hz.
- P300 potentials were recorded using a computerized auditory evoked potential system, calibrated

according to American National Standards Institute standards.

- P300 Latency: Measured as the time (in milliseconds) from stimulus onset to the largest positive peak occurring between 250–500 ms after the target stimulus.
- P300 Amplitude: Measured (in microvolts, μ V) as the voltage difference between the baseline and the peak of the P300 waveform at the Cz electrode.

Hearing Loss Classification

According to World Health Organization (WHO) guidelines, hearing loss was categorized as mild (26 – 40 dB HL), moderate (41 – 55 dB HL), moderately severe (56 – 70 dB HL), severe (71 – 90 dB HL), or profound (> 90 dB HL) ⁽⁵⁾.

Ethical considerations:

All participants or their legal guardians provided written informed permission. The study was approved by Research Ethical Committee, General Organization for Teaching Hospitals and Institutes (GOTHI HAH00066). The Helsinki Declaration was followed throughout the study's conduct.

Statistical analysis

The collected data were coded, entered, and analyzed using the Statistical Package for Social Sciences (SPSS) software, version 27 (IBM, 2020). Quantitative variables were tested for normality using the Shapiro–Wilk test. Quantitative data that followed a parametric distribution were expressed as mean $\pm$ standard deviation (SD), and comparisons among the four study groups were performed using One-way ANOVA (F-test) followed by post-hoc pairwise comparisons (Tukey's test). Qualitative variables were presented as frequency and percentage. Comparisons between groups for categorical data were performed using the Chi-square test (χ^2) or Fisher's exact test when the expected cell count was <5. The significance level was set at $p < 0.05$.

RESULTS

While there was no significant gender difference between the categories, adults with SCT were significantly older than the other groups, the groups' genotypes differed significantly from one another (Table 1).

Table 1. Patient Demographics and Risk Factors in the Study Groups

Parameter	Category	Children with SCD (n=30)	Adults with SCD (n=30)	Children with SCT (n=30)	Adults with SCT (n=30)	Test Results	Post hoc
Age (years)	Mean ± SD	9.87 ± 1.60	31.83 ± 3.40	11.16 ± 0.92	34.95 ± 3.11	F: 858.76, p<0.001*	P1 <0.001 P2 <0.001 P3 <0.001 P4 <0.001 P5 = 0.139 P6 <0.001
Gender	Female	17 (56.7%)	10 (33.3%)	15 (50.0%)	13 (43.3%)	X ² : 3.592, p=0.309	
	Male	13 (43.3%)	20 (66.7%)	15 (50.0%)	17 (56.7%)		
Genotype	HbSC	12 (40.0%)	7 (23.3%)	0 (0.0%)	0 (0.0%)	X ² : 123.983, p<0.001*	P1 <0.001 P2 <0.001 P3 <0.001 P4 <0.001 P5 <0.001 P6 <0.001
	HbSS	16 (53.3%)	21 (70.0%)	0 (0.0%)	0 (0.0%)		
	HbSβ ^o	2 (6.7%)	2 (6.7%)	0 (0.0%)	0 (0.0%)		
	HbAS	0 (0.0%)	0 (0.0%)	30 (100.0%)	30 (100.0%)		

P1: Children with SCD vs Adults with SCD, **P2:** Children with SCD vs Children with SCT, **P3:** Children with SCD vs Adults with SCT, **P4:** Adults with SCD vs Children with SCT, **P5:** Adults with SCD vs Adults with SCT, **P6:** Children with SCT vs Adults with SCT.

There was significant difference between groups regarding hearing loss that was higher in adults with SCD than the three studied groups. Post-hoc analysis revealed that adults with SCD showed a significantly higher rate of hearing loss compared to children with SCT ($p = 0.021$), while other pairwise comparisons were not statistically significant (Table 2).

Table 2. Hearing Loss Assessment: Physical Loss in the Study Groups

Parameter	Category	Children with SCD (n=30)	Adults with SCD (n=30)	Children with SCT (n=30)	Adults with SCT (n=30)	Test Results	Post hoc
No	n(%)	23 (76.7%)	20 (66.7%)	28 (93.3%)	27 (90%)	X ² : 9.128, p=0.028	P1=0.567 P2=0.146 P3=0.299 P4=0.021 P5=0.057 P6=0.99
Hearing Loss	n(%)	7 (23.3%)	10 (33.3%)	2 (6.7%)	3 (10%)		

P1: Children with SCD vs Adults with SCD, **P2:** Children with SCD vs Children with SCT, **P3:** Children with SCD vs Adults with SCT, **P4:** Adults with SCD vs Children with SCT, **P5:** Adults with SCD vs Adults with SCT, **P6:** Children with SCT vs Adults with SCT.

There was no significant difference between the 4 groups regarding hearing loss severity. There was a significant difference between both groups regarding Pure Tone Audiometry (PTA), which was significantly higher in children with SCD than other groups. Pure tone audiometry thresholds were significantly higher in both children and adults with SCD compared to both children and adults with SCT ($p < 0.001$), while no significant difference was observed between children and adults with SCD ($p = 0.153$) (Table 3).

Table 3. Hearing Loss Assessment: Severity and Audiometry in the Study Groups

Parameter	Category	Children with SCD (n=30)	Adults with SCD (n=30)	Children with SCT (n=30)	Adults with SCT (n=30)	Test Results	Post hoc
Hearing Loss Severity	Normal	23 (76.7%)	20 (66.7%)	28 (93.3%)	27 (90%)	X ² : 13.94, P= 0.12	
	Mild	5 (16.7%)	4 (13.3%)	2 (6.7%)	2 (6.7%)		
	Moderate	1 (3.3%)	4 (13.3%)	0 (0.0%)	1 (3.3%)		
	severe	1 (3.3%)	2 (6.7%)	0 (0.0%)	0 (0.0%)		
Pure Tone Audiometry (dB HL)	Mean ± SD	30.20 ± 3.36	28.83 ± 3.95	18.13 ± 1.41	20.20 ± 1.32	F: 143.95, p<0.001*	P1=0.153 P2<0.001 P3<0.001 P4<0.001 P5<0.001 P6<0.001

P1: Children with SCD vs Adults with SCD, **P2:** Children with SCD vs Children with SCT, **P3:** Children with SCD vs Adults with SCT, **P4:** Adults with SCD vs Children with SCT, **P5:** Adults with SCD vs Adults with SCT, **P6:** Children with SCT vs Adults with SCT.

There was no significant difference between groups regarding tympanometry (Type A) and tympanometry (Type B) (Table 4).

Table 4. Tympanometry Results in the Study Groups

Parameter	Category	Children with SCD (n=30)	Adults with SCD (n=30)	Children with SCT (n=30)	Adults with SCT (n=30)	Test Results
Tympanometry (Type A)	n(%)	26 (86.7%)	30 (100.0%)	28 (93.3%)	29 (96.7%)	X ² : 5.310, p=0.150
Tympanometry (Type B)	n(%)	4 (13.3%)	0 (0.0%)	2 (6.7%)	1 (3.3%)	X ² : 5.310, p=0.150

IQ was significantly lower in children with SCD than other groups. There was a significant difference between groups regarding cognitive dysfunction that was extremely low in children and adults with SCD than other groups. Post-hoc analysis demonstrated that cognitive dysfunction was significantly more prevalent among both children and adults with SCD when compared with SCT groups ($p < 0.001$). No significant difference was observed between the SCT children and adults ($p = 0.312$) (Table 5).

Table 5. Cognitive assessment in the Study Groups

Parameter	Category	Children with SCD (n=30)	Adults with SCD (n=30)	Children with SCT (n=30)	Adults with SCT (n=30)	Test Results	Post hoc
IQ	Mean $\pm$ SD	78.98 $\pm$ 10.04	86.28 $\pm$ 11.61	90.94 $\pm$ 5.50	90.70 $\pm$ 4.39	F: 13.15, p<0.001*	P1=0.012 P2<0.001 P3<0.001 P4=0.054 P5=0.059 P6=0.853.
Cognitive Dysfunction	Borderline	12 (40.0%)	8 (26.7%)	0 (0.0%)	0 (0.0%)	X ² : 44.502, p<0.001*	P1=0.044 P2<0.001 P3<0.001 P4=0.002 P5=0.001 P6=0.312
	Extremely Low	5 (16.7%)	2 (6.7%)	0 (0.0%)	0 (0.0%)		
	Low Average	9 (30.0%)	8 (26.7%)	15 (50.0%)	10 (33.3%)		
	Average	4 (13.3%)	12 (40.0%)	15 (50.0%)	20 (66.7%)		

P1: Children with SCD vs Adults with SCD, **P2:** Children with SCD vs Children with SCT, **P3:** Children with SCD vs Adults with SCT, **P4:** Adults with SCD vs Children with SCT, **P5:** Adults with SCD vs Adults with SCT, **P6:** Children with SCT vs Adults with SCT.

P300 latency was significantly higher in children with SCD than other groups and P300 Amplitude was significantly higher in children with SCT than other groups. P300 latency was significantly prolonged and P300 amplitude was significantly reduced in both children and adults with SCD compared to SCT groups ($p < 0.001$). No significant difference in P300 parameters was found between children and adults with SCD regarding amplitude ($p = 0.533$), whereas latency remained significantly different ($p < 0.001$) (Table 6).

Table 6. Neurological assessment in the Study Groups

Parameter	Category	Children with SCD (n=30)	Adults with SCD (n=30)	Children with SCT (n=30)	Adults with SCT (n=30)	Test Results	Post hoc
P300 Latency (ms)	Mean ± SD	439.17 ± 10.47	426.20 ± 9.22	375.10 ± 3.56	389.13 ± 8.17	F: 399.90, p<0.001*	P1 <0.001
							P2 <0.001
							P3 <0.001
							P4 <0.001
							P5 <0.001
							P6 <0.001
P300 Amplitude (mV)	Mean ± SD	12.27 ± 1.53	12.03 ± 1.43	16.97 ± 0.76	14.93 ± 0.87	F: 115.73, p<0.001*	P1=0.533
							P2 <0.001
							P3 <0.001
							P4 <0.001
							P5 <0.001
							P6 <0.001

P1: Children with SCD vs Adults with SCD, **P2:** Children with SCD vs Children with SCT, **P3:** Children with SCD vs Adults with SCT, **P4:** Adults with SCD vs Children with SCT, **P5:** Adults with SCD vs Adults with SCT, **P6:** Children with SCT vs Adults with SCT.

DISCUSSION

The demographic information in our study showed notable age ranges within the study groups, with the mean ages of the adults and children ranging from 31.83 to 34.95 years and 9.87 to 11.16 years, respectively. The gender distribution revealed that adult SCD patients had more men (66.7%), while children with SCD had a small female predominance (56.7%).

This is in line with the findings of **Youssry et al.** ⁽⁶⁾, who sought to evaluate the cognitive and auditory abilities of SCD patients. They stated that the children with SCD ranged in age from 6 to 17 years (mean± SD = 10.4±3.3 years), with 26 (63.4%) being females and 15 (36.6%) being males.

Al Jabr ⁽⁷⁾ sought to determine the prevalence of sensorineural hearing loss in sickle cell disease patients in adults. They stated that the patient group's age ranged from 20 to 45 years old and a mean (standard deviation) of 31.7 (6.5) with a male to female ratio of 1.9:1.

The evaluation of hearing loss severity in our study showed no statistically significant difference between the SCD and SCT groups. A modest proportion of SCD patients exhibited hearing loss (approximately 23.3% in children and 33.3% in adults), whereas most SCT patients maintained normal hearing (93.3% in children and 90% in adults). Correspondingly, the Pure Tone Audiometry (PTA) mean values were 30.20 ± 3.36 dB in children with SCD, 28.83 ± 3.95 dB in adults with SCD, compared with 18.13 ± 1.41 dB in children with SCT and 20.20 ± 1.32 dB in adults with SCT.

These results are consistent with a study by **Miri-Aliabad et al.** ⁽⁸⁾, which likewise discovered that SCD patients had more hearing loss than control groups.

Pure tone audiometry (PTA) is the gold standard test and the best choice for screening peripheral hearing loss when a child's hearing threshold is less than or equal to 20 dB ⁽⁹⁾.

Accordingly, **Ibekwe et al.** ⁽⁴⁾ found that 30% of adults with SCD, 36.1% with SCT, and 11% with HbAA had hearing impairment, compared to 35% of children with SCD and 25% with SCT. Children's left ear was more impacted than adults' right, indicating asymmetry in the hearing impairment.

Youssry et al. ⁽⁶⁾ revealed that 22% of children with SCD had hearing loss overall, as determined by pure tone audiometry, and that over 75% of patients—mostly those with the Hb SS subtype—had cognitive impairment that worsened with age.

According to **Al Jabr** ⁽⁷⁾, nine patients (22.5%) received a pure tone audiometry diagnosis of hearing loss, despite the fact that only five patients (12.5%) among adults with SCD complained clinically of hearing loss. Additionally, there was a statistically significant difference in the means of the hearing thresholds on all frequencies between the patients and the controls.

The Centers for Disease Control and Prevention found that among children aged 3–17 years who were chosen for participation in a nationwide survey without consideration for symptoms, the parent-reported prevalence of hearing loss was 0.5% ⁽¹⁰⁾. Based on the average of eight similar studies that used PTAs for audiometric examinations in children aged 3 to 19, a systematic review calculated the prevalence of hearing impairment to be 3.1% ⁽¹¹⁾. Children aged 6-19 years had a prevalence of up to 14.9%, according to the Third National Health and Nutrition Examination Survey ⁽¹²⁾.

Worldwide, reports of the prevalence of hearing loss in SCD patients, independent of the type, ranged from 20% ⁽¹³⁾ to 24% ⁽¹⁴⁾, with approximately identical frequencies. In contrast, a prior study conducted in the USA ⁽¹⁵⁾ revealed a frequency of 0 percent; in **Onakoya et al.** ⁽¹⁶⁾, the incidence was substantially greater, ranging from 45.6% to 66%.

According to the results of our tympanometry, type A tympanograms were more common in all categories, but they were especially prevalent in adults with SCD (100%) and SCT (96.7%). Children with SCD had a significantly greater prevalence of type B tympanograms (13.3%), which may suggest that they are more likely to have middle ear effusions.

Youssry et al. ⁽⁶⁾ evaluated auditory functions and discovered that 5% of patients had Eustachian tube dysfunction (Type C tympanogram) and nearly 10% of patients had middle ear effusion (Type B tympanogram), which are signs of middle ear pathology.

In contrast to a study conducted in the early 1980s that found that 34% out of 54 Hb SS children, aged 12-years, had abnormal tympanometric results (14% with type B and 20% with type C) ⁽¹⁷⁾, the frequency of type B tympanogram pathology was significantly lower among their children.

According to **Piltcher et al.** ⁽¹⁸⁾, six out of 28 (21%) SCD patients, ages 6 to 55, had defective tympanometry. The exclusion of certain confounding factors, such as chronic otitis media or prior exposure to ototoxic medications, or a real decline in otitis media rates brought on by widespread vaccination against *Hemophilus influenzae*, reduced exposure to ototoxic drugs, and timely treatment of vaso-occlusive crises, may account for the lower percentage.

The results of our study's cognitive evaluation showed that the groups' IQ scores varied significantly. Compared to both adult SCD patients (86.28 ± 11.61) and SCT groups (around 90 for both children and adults), children with SCD had lower mean IQ scores (78.98 ± 10.04). With 16.7% exhibiting severely low cognitive function and 40% exhibiting borderline function, the distribution of cognitive dysfunction in children with SCD was especially worrisome.

This result is consistent with studies by **Salama et al.** ⁽¹⁹⁾ that found impaired cognitive effects in children with sickle cell disease.

According to **Youssry et al.** ⁽⁶⁾, up to 80.5% of children with SCD had cognitive impairment, and over 40% of all patients were slow learners with 76 was the median score for global cognitive IQ. This was comparable to findings from earlier research that showed mean scores ranging from 75.6 to 82 ⁽²⁰⁾. Other investigations have recorded higher scores, up to 101 ⁽²⁰⁾.

Youssry et al. ⁽⁶⁾ reported that the absence of fetal hemoglobin, which guarantees improved brain

oxygenation, makes patients with the Hb SS genotype more vulnerable to disease-related consequences. They may experience silent infarctions and irregularities in the structure of their brains, which increase their susceptibility to alterations in brain perfusion.

Variations in reported scores could be due to differences in ethnicity, disease severity, or care between centers, which could result in hidden brain damages that impair intellectual capacity.

Through P300 measures, the neurological evaluation in our study objectively demonstrated variations in cognitive processing. In comparison to SCT groups (375.10 ± 3.56 ms in children and 389.13 ± 8.17 ms in adults), SCD patients exhibited noticeably delayed P300 latencies (439.17 ± 10.47 ms in children and 426.20 ± 9.22 ms in adults). Furthermore, SCD groups showed a significantly lower P300 amplitude, indicating a diminished capacity for cognitive processing.

These results are consistent with neurophysiological research by **Colombatti et al.** ⁽²¹⁾ that found P300 aberrations in SCD patients to be comparable.

The P300 is an auditory evoked potential that can assess cognitive abilities such as memory, sound discrimination, and attention level. It is a measure of speed recognition and cortex processing ⁽²²⁾.

According to **Youssry et al.** ⁽⁶⁾, 29.3% of the studied patients had excessively lengthy P300 latency. The absence of correlation between P300 latency and all disease severity parameters points to a silent disease-specific alteration in the affected patients' cerebral cortex. This could be connected to the chronic hypoxia brought on by persistently low hemoglobin or the ongoing inflammatory response in SCD patients. Additionally, the absence of association with silent infarction may indicate broad brain injury leading to neuronal impairment. Because fetal hemoglobin (Hb F) keeps red blood cells from sickling, more oxygen is distributed throughout the body, including the brain. The most important laboratory metric for determining the clinical severity of sickle cell disease is Hb F.

Sadly, not much is known about P300 test results in children with sickle cell disease ⁽²¹⁾. However, **de Castro et al.** ⁽¹³⁾ found that both groups' P300 amplitude and latency were sufficient, indicating that there were no anomalies in the central auditory system. According to their research, SCD causes varying degrees of cochlear defects without demonstrating any neurological issues.

CONCLUSION

The present study demonstrated that sickle cell disease (SCD) exerts a more profound impact on auditory, cognitive, and neurophysiological functions than sickle cell trait (SCT), particularly among children. Although the overall prevalence and severity of hearing loss were not statistically significant between the SCD and SCT

groups, SCD patients exhibited higher mean pure tone audiometry (PTA) thresholds, indicating subtle cochlear involvement. Tympanometric findings further suggested an increased risk of eustachian tube dysfunction in SCD children. Moreover, both IQ assessment and P300 measurements revealed considerable cognitive and central processing delays in SCD patients, highlighting a systemic neuro-auditory vulnerability. These findings emphasize the importance of early audiological screening, neurocognitive monitoring, and timely interventions in individuals with SCD to mitigate long-term developmental and quality-of-life impairments. Future studies with larger sample sizes and longitudinal follow-up are warranted to elucidate the underlying mechanisms and to evaluate the effectiveness of early therapeutic and rehabilitative strategies.

Funding: No fund.

Availability of data & material: Available.

Conflicts of interest: None.

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