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Prevalence of hearing loss in patients with sickle cell disease in the southeast of Iran

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Abstract

Background: One of the complications of sickle cell disease (SCD) is hearing loss. The purpose of the present study is to determine the prevalence of hearing loss in SCD.

Materials and Methods: This is a descriptive-analytical cross-sectional study conducted on 100 patients with SCD in 2019-2020. All the patients underwent otololaryngological examination and audiometric tests to assess their hearing status. The type and severity of hearing loss were determined, the demographic information of the patients was recorded, and the data were analyzed using the SPSS software version 21, a Chi-square test, and the Kruskal-Wallis test. The P-values of < 0.05 were considered statistically significant.

Results: The mean age of the patients was 17.6 ± 10.8 years. Of them, 60% were male. Hearing loss was diagnosed in 26 patients (26%), 16 of whom had sensorineural hearing loss (SNHL) and 10 had conductive hearing loss (CHL). In terms of the disease severity, 12, 10, 3 and 1 patients had mild, moderate, severe and profound hearing loss, respectively. The mean age of the patients with SNHL was significantly higher than the median age of the subjects with CHL (p-value = 0.043). Also, the patients with hearing loss had no statistically significant difference in terms of the variables of age, gender, unilateral or bilateral ear involvement and type of SCD (p-value = 0.069).

Conclusion: This study revealed a hearing loss frequency of 26% among patients with SCD. Regular hearing assessment and timely treatment and rehabilitation measures are recommended for these patients.

Keywords: Hearing loss, Prevalence, Sickle cell disease

Introduction

Sickle cell disease (SCD) is one of the most prevalent hereditary disorders worldwide, characterized by hemoglobin S in erythrocytes. This results in the production of sickle-shaped red blood cells, which are less flexible and smaller than normal red blood cells (1). Veno-occlusive disease, hemolytic anemia, ischemia and tissue damage are the obvious signs of SCD (1, 2). In SCD, many organs such as the spleen, bones, lungs, kidneys, and nervous system are affected (3). The brain is also affected both structurally and functionally, which predisposes children to cognitive impairment, transient ischemic attacks, and strokes (3, 4). In patients with SCD, the cochlea is affected too, leading to hearing loss (5). The incidence of hearing loss in different populations of patients with SCD has been reported to be different; in some studies, no hearing loss was observed but, in some others, even 66% of the population had that hearing disorder(1). Hearing loss and the resulting speech disorders cause many problems in educational and social fields (6). There is no program for the regular periodic evaluation of children with SCD. Early detection of hearing impairments can improve the effectiveness of therapeutic interventions and educational performance (6).

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The pathological changes of SCD are caused by vaso-occlusion, chronic hemolytic anemia and infection.

The most common pathological factor causing hearing loss and the involvement of the inner ear in SCD is recurrent vaso-occlusion in the labyrinth (7-10). Over the last three decades, few studies have examined hearing impairment in children with SCD (11-16). The research in this field has mainly been on a combination of adults and children. This has led to a lack of understanding about how risky hearing impairment is in children alone (16, 17). In addition, many studies have been conducted in countries where sensorineural hearing loss (SNHL) is common due to endemic infections such as bacterial meningitis, malaria, and congenital rubella (18-20). Also, most studies have only examined limited SNHL. In a study by Al Jabr et al. (21), 22.5% of the SCD patients had hearing loss. Their hearing threshold in both ears at all frequencies was significantly different from that in the control group. In this context, the present study seeks to determine the frequency of different types of hearing impairment in subjects with SCD.

Materials and Methods

This study is a cross-sectional descriptive-analytical one. The study population included 100 subjects with SCD who referred to the hematology clinic of Ali Asghar Children's Hospital in 2019-2020. The diagnosis of SCD was confirmed through hemoglobin electrophoresis and/or genetic analysis. Initially, the patients were subjected to physical examination, history inquiry, and detailed evaluation by a hematologist, an otolaryngologist and an audiologist. The exclusion criteria included a history of chronic otologic problems, otologic surgery, exposure to excessive noise or ototoxic drugs, previous hearing loss due to trauma, meningitis, mumps, measles and rubella. The demographic information of the patients was recorded, and their speech disorders were assessed. The audiologic assessment was performed by an audiologist using an interacoustic audiometer model AC 33 and a Madsen tympanometer device model ZODIAC 901. To assess the conditions of the outer and middle ears, otoscopic examination, tympanometry test and stapedius reflex test were performed. All the subjects underwent pure tone audiometry (PTA). The pure tone thresholds higher than 25 dB in one or more test frequencies from 500 to 4000 Hz were considered as hearing loss. The audiograms were also analyzed into conductive, sensorineural and mixed domains of hearing loss. Hearing loss was categorized into mild (threshold of 26-40 dB), moderate (41-60 dB), severe (61-80 dB), and profound (> 80 dB).

Statistical analysis

The SPSS software version 21 served to analyze the data. This study not only used descriptive statistical indices including frequency, percentage, mean, variance and standard deviation but also benefited from a Chi-square test, an independent t-test, and the one-way analysis of variance. A P-value less than 0.05 was considered significant.

Results

The present study was conducted on 100 patients with SCD aged 6 to 45 years. The mean age of the subjects was 17.6 ± 10.8 years. Of the patients, 40 (40%) were female, and 60 (60%) were male. The results of the study showed that, in all the age groups, less than one third of the patients had hearing loss and more than two thirds of them were normal in terms of hearing loss. In this regard, there was no statistically significant difference among the age groups (p = 0.689). Also, 25% of the females and 26.7% of the males had hearing loss (p = 0.852) (Table I).

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Based on Bonferroni test, the age difference between the two groups of SNHL and conductive hearing loss was found to be significant (p-value = 0.043). However, no significant age difference was found between any of these two groups and the normal group (p–values of 0.352 and 0.297, respectively) (Figure 1).

In the present study, 10 patients received iron chelators, of whom only two had hearing loss. There was no significant difference in hearing loss between the people who used iron chelators and those who did not (P = 0.648).

Table II shows the types of hearing loss in the patients with SCD. Eleven of them had unilateral and 15 had bilateral ear involvement. Also, 81.8% of those with unilateral ear involvement had SNHL. There was no significant difference in the prevalence of the unilateral and bilateral types of hearing loss (p = 0.069).

Table I: Frequency distribution of different types of hearing loss in patients with SCD by age group and gender

Variable		Hearing loss	Normal hearing	P-value
Age group (years) No (%)	< 10	7 (21.9)	25 (78.1)	0.689
	11-20	10 (32.3)	21 (67.7)	
	21-30	6 (21.4)	22 (78.6)	
	> 30	3 (33.3)	6 (66.7)	
Gender No (%)	Male	10 (25)	30 (75)	0.852
	Female	16 (26.7)	44 (73.3)	

Table II: Frequency distribution of hearing loss in patients with SCD according to unilateral or bilateral ear involvement

Ear involvement	SNHL	CHL	Total, No (%)	P-value
Unilateral	9 (81.8)	2 (18.2)	11 (100)	0.069
Bilateral	7 (46.7)	8 (53.3)	15 (100)	
Total, No (%)	16 (61.5)	10 (38.5)	26 (100)	

Abbreviations: SNHL: sensorineural hearing loss, CHL: conductive hearing loss

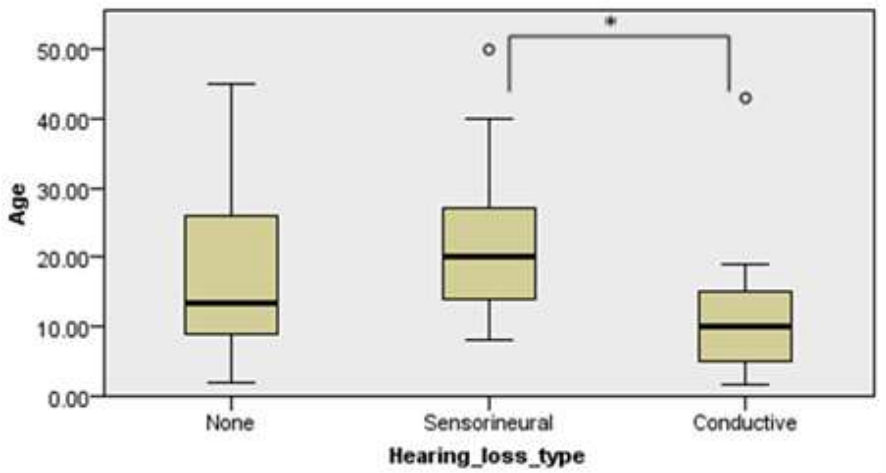

Figure 1. Means and standard deviation of age for different types of hearing loss in patients with SCD

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Discussion

Of the SCD patients participating in this study, 26 (26%) had hearing loss. Sixteen cases of the loss were SNHL and 10 were conductive. Based on the study of Rissatto Lago et al. (16), who reviewed some previous studies, the rate of SNHL in patients with SCD is higher than that in healthy individuals. In this regard, there is a consensus among most reports. Different patterns and degrees of hearing loss have also been reported in different studies. They range from profound and bilateral hearing loss with relative improvement over time to mild unilateral or bilateral hearing loss with higher frequency (22). In the research by Stuart et al. (1), the frequency of hearing impairment in homozygous sickle cell patients was reported to be 28.8-50.8%. In the present study, however, a different range of hearing loss was observed; among 26 patients with hearing loss, the highest severity was related to mild (46.2%) and moderate (38.5%) levels of hearing loss.

A review study in 2019 (16) showed that patients with SCD may experience hearing changes of cochlear or retro-cochlear origin. SNHL was also found more common in patients with SCD (20.5%) than in healthy individuals (0 to 7.5%). Moreover, it was concluded that SNHL can develop at different ages, as in children, adolescents, and young adults. The results of that study are consistent with the present study. It seems that the main reason for SNHL in people with SCD is cochlear damage caused by circulatory disorders, which induce chronic inflammatory conditions (16). Several pathophysiological factors have been proposed for hearing impairment in sickle cell anemia patients. CHL usually occurs secondary to middle ear disorders following upper respiratory infections. Adenotonsillar hypertrophy and functional asplenia in SCD patients lead to increased susceptibility to infections and otitis media, which may be the cause of conductive hearing loss (23). Damage to the endothelium of vessels, inflammation, or the sickling of red blood cells leads to decreased blood flow, impaired blood supply of the cochlear venous system, hypoxia of the Organ of Corti and, ultimately, sensorineural hearing impairment (15). Schopper et al. (24) found that 62% of the patients with SCD had hearing loss, and higher intensities of hearing loss were more visible. These findings are inconsistent with the results of the present study and some similar studies that have shown hearing loss with the prevalence of about 20 to 30% and the highest severity of the loss in milder degrees. A possible explanation for the inconsistency of these findings is the differences in the methods of implementing the plans, such as how to select samples, or even the effect of some genetic factors and diagnostic therapeutic protocols, especially since the main hearing loss in that study was seen in blacks. In a study on 89 children with SCD, Bois et al. (4) found that 7.1% of the patients with normal hearing had speech disorders. Almost similarly, in the present study, 74% of the patients had normal hearing, of whom 8 (10.8%) suffered from speech disorders without hearing loss. Speech disorders in these patients can have different causes. A study by Lieu JE showed that unilateral hearing loss can affect the educational status of children and cause speech and language disorders (25). Patients with SCD without screening and prophylactic treatment have an increased risk of stroke; as reported, the prevalence of first stroke in children and adolescents with SCD is in the range of 5-17% and the life time risk of overt stroke is 25-30% (26). In a study by Al Okbi et al. (2) in Oman, SNHL was examined in patients with SCD, and ten hearing-impaired patients were found with simultaneous bilateral ear involvement and

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four with unilateral involvement. In a research by Towerman et al. (27), 19.4% of the patients with SCD had hearing loss, which was slightly less common than the finding of this study. In general, the literature indicates a higher frequency of bilateral ear involvement than unilateral involvement in patients with SCD, which is consistent with the results of the present study.

Conclusion

The range of hearing impairment in this study varied from normal to severe hearing loss in both conductive and SNHL types. This disorder can be unilateral or bilateral. The severity of hearing loss in each ear can also vary. In the present study, 26% of the patients had hearing loss. Due to the high percentage of hearing loss among these patients, hearing tests at regular intervals from the time of diagnosis seem necessary for the early detection of hearing impairment, timely treatment, and rehabilitation interventions.

Ethical considerations

Informed consent was obtained from all the patients. The study was also approved by the University Ethics Committee (Ethics code: IR.ZAUMS.REC.1398.362).

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Conflict of Interests

The authors had no conflict of interests to declare.

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Author`s Contribution

Ghasem Miri-Aliabad: Study concept, study design, manuscript writing and literature search.
Majid Naderi: Manuscript editing, literature search.
Aziz Eghbali: Manuscript editing, literature search.
Ebrahim Pirasteh: Statistical analysis, manuscript editing.
Farzad Hamzehpour: Data acquisition, audiometry.
Asma Erish: Data acquisition, manuscript writing.

Conflict of interest

The authors declare no conflict of interest.

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