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A STUDY OF SENSORINEURAL HEARING LOSS IN CHILDREN AFFECTED WITH SICKLE CELL DISEASE

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ABSTRACT

Background: Sickle cell disease (SCD) is one of the common genetic disease in India. It is often associated with a high incidence of hearing loss which generally goes undiagnosed in majority of children . Our study aims to determine the prevalence of sensorineural hearing loss in children of Chhattisgarh with SCD in order to provide a close audiological follow-up and sensitise medical professionals for early Detection and rehabilitation of SNHL in children with SCD.

Methods: This study included 280 children of 0-18 years of age group with and without complaint of hearing loss. Clinical examination of the children was done including general examination and examination of ear, nose and throat. The Pure Tone Audiometry (PTA) was done by an audiometrist in a sound proof room using a calibrated audiometer "Interacoustic AC 40" at frequencies ranging from 250 Hz to 8000 Hz. Otoacoustic Emissions (OAE) and BERA was conducted by GSI (Grason Stadler) Audera machine.

Results: Among 280 patients 9 (3.2%) patients presented with sensorineural hearing loss. The incidence of hearing loss was higher (55%) in males than in females (44%) and it was more frequent in the 10-15 yrs. of age group. All 9 cases with hearing loss were homozygous for sickle cell disease and majority of them (4/9) were suffering from severe anemia. Conclusions: The overall consequences of sickle cell disease on hearing and auditory function is significant and cannot be overlooked. As the mortality and morbidity associated with sickle cell anaemia has decreased, management of associated health risks such as hearing impairment will greatly benefit from early diagnosis and rehabilitation.

KEYWORDS

sensorineural hearing loss, prevention, sickle cell disease, children,

INTRODUCTION

It is now a recognized fact, that as earlier the deafness is detected in childhood, better are the prospects of a successful education. Due to lack of awareness and knowledge about hearing loss in children it often goes undiagnosed, and when ignored in childhood or treated incompletely may result in permanent impairment of speech and hearing.

There are several reports indicating association of Sickle cell anaemia and sensorineural hearing loss (SNHL) in children. Sickle cell anaemia is widely distributed in many parts of the world. It is a genetically inherited haemoglobinopathy in which the amino acid Valine is substituted for Glutamic acid in the sixth position of the beta chain of haemoglobin. The existence of this entity had been recognized in Africa in the 20th century. Most of these evidences came from Ghana.[5] It was for the first time reported in India by Lehman and Cutbush (1952)[6] amongst the tribes of Nilgiri Hills in South India. In Chhattisgarh region Gupta and Tiwari[7] described 30.25% and 17.7% incidence of sickle cell anaemia respectively amongst tribal population. Todd et al [8] in their study reported that sensorineural hearing loss (SNHL) in 22% cases of homozygous sickle cell anaemia in 83 patients aged 10-39 years. Another study by Oyebanji Anthony Olajuyi et[9] al on the otological burden of sickle cell disease (SCD) in 168 Nigerian children at a tertiary health care centre in Nigeria was conducted suggested that both the hemoglobin concentration and HbF did not discriminate between the SCD participants with or without SNHL. This study showed that otological burdens are more prevalent in children with SCD than the non-SCD population.

In another study conducted by Hunter et al [10], the increased Distortion Product Oto Acoustic Emissions (DPOAEs) amplitude was believed to be an early sign and precursor before the onset of hearing loss, which was evident in children with SCD not on Hydroxyurea. Those taking Hydroxyurea had DPOAE amplitudes similar to normal controls.

MATERIALS AND METHODS

The study was conducted on 280 patients of both sexes in the age group of 0-18 years with and without complaint of hearing loss, visiting the outpatient Departments of ENT, Pediatrics and Medicine of Dr. BRAM Hospital as well as the Sickle Cell Research Institute, Raipur.

Permission from the institutional scientific and ethical committee was taken and all participants were informed regarding the study and written consent was obtained. Detailed history and clinical examination was carried out in all the children and then they were subjected to audiological evaluation by Pure Tone Audiometry, OAE and BERA for detection of hearing loss.

The Pure Tone Audiometry (PTA) was done by an audiometrist in a sound proof room using a calibrated audiometer "Interacoustic AC 40" at frequencies of 250 Hz, 500 Hz, 1000 Hz, 2000 Hz, 4000 Hz and 8000 Hz. Otoacoustic Emissions (OAE) and BERA was conducted by GSI (Grason Stadler) Audera machine. Hearing loss was considered significant if a child had an average hearing threshold of more than 25 dB at two or more frequencies by PTA in one ear.

The inclusion criteria was children positive for sickle cell disease (SS) or trait (AS) in the age group of 0-18 years while the exclusion criteria included patients with or without hearing loss in the same age group but not suffering from sickle cell anaemia.

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RESULTS

This study was undertaken on 280 patients of sickle cell disease including 149 (53.2%) males and 131 (46.7%) females of age group

ranging from 0-18 years. The age and gender distribution of patients has been shown in **Table 1**.

S.No	Age Group in yrs	Male No. (%)	Female No. (%)	Total No. (%)
1	4-6	13 (4.64)	12 (4.28)	25 (8.92)
2	7-9	42 (15.0)	29 (10.35)	71 (25.35)
3	10-12	30 (10.71)	36 (2.85)	66 (13.56)
4	13-15	28 (10.0)	17 (6.07)	45 (17.07)
5	16-18	36 (12.85)	37 (13.21)	73 (26.06)
Total		149 (53.21)	131 (46.79)	280 (100)

Among these 9 (3.2%) patients presented with sensorineural hearing loss. The incidence of hearing loss was higher (55%) in males than in females (44%) and it was more frequent in the 10-15 yrs of age group. Among the 280 cases, 277 patients had centralized Weber's test, 2 patients had lateralization of the Weber towards right while 1 patient had lateralization to the left. The absolute bone conduction test was equal in 271 while reduced in 9 patients. The Pure Tone Audiometry (PTA) and Brainstem Evoked Response Audiometry (BERA) findings revealed that out of 9 hearing loss cases 5 patients showed mild (30 dB), 3 moderate (50 dB) and 1 severe (80dB) degree of hearing loss. Similarly, all patients with hearing loss showed absence of Distortion Product Oto Acoustic Emissions (DPOAE) as shown in **Table II** depicting DPOAE Findings of patients**.**

Table II DPOAE Findings Of Patients (N=280)

Test	Present	%	Absent	%
DPOAE	271	97	9	3

The above table shows that DPOAE were present in 271 out of 280 patients (97%) and absent in 9 out of 280 patients (3%).

In majority (8/90) of hearing loss cases higher frequency (4000-8000 Hz) was involved. It was further observed that the subjects with early onset of vascular crisis the degree of hearing loss was found to be severe as shown in **Table III**.

Table III Age At First Vascular Crisis In Patients With Hearing Loss.

S. No	Case No.	Age at first vascular crisis	PTA in dB Rt	Lt	Degree of SNHL
1	Case 1	6	32.5	33.7	Mild
2	Case 2	8	37.5	37.5	Mild
3	Case 3	5	31.2	27.5	Mild
4	Case 4	8	45	46.2	Moderate
5	Case 5	6	45	47.5	Moderate
6	Case 6	6	80	73.3	Severe
7	Case 7	0	35	35	Mild
8	Case 8	5	31.2	32.5	Mild
9	Case 9	12	51.2	50	Moderate

All 9 cases with hearing loss were homozygous for sickle cell disease and majority of them (4/9) were suffering from severe anaemia (<7gm/dl according to WHO criteria). However, no correlation was found between the degree of hearing loss and level of anemia.

DISCUSSION

Sickle cell anaemia is a genetically determined condition characterized by genetic mutation and presence of abnormal haemoglobin. The systemic manifestations of this disease develop as a result of vascular occlusion caused by sickle shaped red blood cells. This gives rise to various signs and symptoms such as abdominal pain, joint pain, chronic and recurring leg ulcers, priapism, pulmonary embolism, cardiac infarctions and various signs related to the central nervous system.

Hearing loss associated with SCD may occur both in children and adults. It can be unilateral or bilateral, mild or severe, transient or permanent and its onset can be sudden or progressive. More often it is due to inner ear pathology rather than external ear or middle ear complications. The hypothesized mechanisms of the SNHL in patients with SCD include impairment of oxygen supply (secondary to deformed RBCs), which results in hypoxia to the cochlea, peripheral and central auditory pathways, sluggish blood flow to the cochlea due to sickling of the red blood cells, and compression of the auditory canal and auditory nerve by the hyperactive bone marrow in the petrous part of the temporal bone. Another possible mechanism of hearing loss in SCD patient is increased susceptibility to bacterial meningitis, as the immune function of these patients is affected.

The concern with the early diagnosis of hearing impairment has been a constant issue, since the loss caused by such impairment, often times is irreversible, affecting not only oral language, but also the child's global development and school performance. In the present study observations made from 280 cases of varying age and sex between 0-18 years in our Institute. Older age groups were not taken due to presence of arteriosclerosis and presbycusis contributing to hearing loss in them. These patients were subjected to clinical examination and audiological evaluation by pure tone audiometry, otoacoustic emissions and Brainstem Evoked Response Audiometry. Out of 280 cases of sickle cell anaemia 9 (3.2 %) subjects showed bilateral sensorineural hearing loss.

The incidence of SNHL in this region appears to be less compared to other part of the country. In another study from Bhatinda, 20 out of 105 (19%) patients were reported to have sensorineural hearing loss by Priyanka et al11 2019. However, in this study both conductive and sensorineural hearing loss were considered, while we assessed only sensorineural hearing loss because in sickle cell disease cases hearing loss is most often due to inner ear pathology (SNHL) than to external ear or middle ear complications (CHL).

In the study conducted by Ibrahim et al[12], 9 out of 40 (22.5%) patients were seen to have hearing loss. The age range taken in this study was 20-45 years. Since as the age increases, the number of episodes of vascular crisis also increases, the older age group might be a reason for more incidence of hearing loss seen in this study as compared to ours. Of the 9 (22.5%) patients who had hearing loss in their study 6 (15%) had bilateral hearing loss while 3 (7.5%) had unilateral hearing loss. In our study all the 9 subjects who had hearing loss, bilateral involvement was seen. In our study, incidence of sensorineural hearing loss was maximum in age group 10-12 years and 13-15 years (33% each) followed by 16-18 years age group (22%). These findings are consistent with possible more attacks of sickle cell crises in older age group subjects.

In our study the hearing loss was almost equally distributed in both sexes (male: female, 5:4) and in both ears. Similar observation were made in study earlier done by Friedman et al.[13]A significant relationship between high frequency sensorineural hearing loss and DPOAE's has been shown by Burch-Sims et al[14]earlier. DPOAE's were consistently present in frequency regions exhibiting normal pure tone sensitivity. DPOAE amplitudes were reduced as pure tone thresholds were increased, and they were absent at frequencies where threshold were greater than 50 dB for 251 out of the 255 ears tested. For four ears, DPOAE and pure tone audiograms were not correlated. In our study also DPOAE and pure tone hearing thresholds were correlated in all cases of hearing loss.

In most of the subjects having hearing loss (8 out of 9), higher frequencies (4000-8000 Hz) are involved more than lower frequencies. This shows that most people with sickle cell anaemia who have hearing loss, have high frequency hearing loss.

This is exemplified by the study of Koide et al [15]and Conti and Borgo[16] who noted that the sensitivity of the basal turn of the cochlea to anoxia was due to high oxygen consumption rate of stria vascularis in this area and to the poor capacity for anaerobic metabolism. The basal turn of the cochlea records high frequencies and the apical turn records low frequencies.Accordingly in experimental venous obstruction it might be expected that high frequencies would be first affected followed by low frequencies and ultimately deterioration in all frequencies.

A sickle cell crisis is a painful episode that may begin suddenly in a person who has sickle cell disease. In our study sickle cell crisis history was present in 48 (17.1%) of the cases and 8/48 (16.6 %) showed hearing loss. In remaining subjects who did not suffer from sickle cell crisis, only 1/48 (2%) subjects had hearing loss. Further, our results show that as the number of episodes of vascular crisis increase, the degree of hearing loss increases (**Table IV**).

Table IV: Case Distribution Of No. Of Episodes Of Vascular Crisis

S no.	Episodes of vascular crisis	Cases	%
1	1	1	11
2	2	3	33

3	3	3	33
4	4	1	11

According to this table, of the total 9 cases experiencing hearing loss, 1 case had 1 episode of vascular crisis, 3 cases has 2 episode, 3 cases had 3 episodes and 1 case had 4 episodes of vascular crisis. 1 case had hearing loss but no history of vascular crisis. In our study, one case of homozygous sickle cell disease (SS) with severe degree of sensorineural hearing loss was found to have a maximum of 4 episodes of vaso-occlusive crisis.

Ischaemia of the stria vascularis, with secondary organ of Corti hypoxia as a direct result of sickle cell crisis, poses significant risk to the auditory system. Increased blood viscosity by crystallization of red blood cells, with subsequent slugging, development of stasis, ischaemia or hypoxia appears to be the outstanding pathophysiological phenomenon during crisis as explained by Diggs et al. [17]

Thus it is obvious that sickle cell crisis affects hearing apparatus almost 2.8 times more and seems to be mainly responsible for hearing loss in sickle cell disorders. As is natural, hearing loss was more in 10-12 years age group with sickle cell crisis. Each sickle cell crisis causes loss of cell membrane and produces irreversible sickle cells in blood. Presence of irreversibly sickled cells in blood increases the blood viscosity leading to haemostasis along with local hypoxemia and acidosis, thereby promoting further sickling and ultimately leading to vaso occlusion.

Our results were found parallel to earlier studies done by Morgenstein and Manace[18], and George E Urban[19] showing high incidence of sensorineural hearing loss in patients with sickle cell crisis. In the study of temporal bone performed by Seargent et al [20] in subjects who had repeated crises, observed that chronic hypoxia in the inner ear was the cause of the structural changes in the outer and inner hair cells throughout the organ of corti leading to sensorineural hearing loss. Our study shows that out of 280 total patients 220 were heterozygous (AS) and 60 were homozygous (SS) for sickle cell disease. Further 2 out of 9 patients with hearing loss suffered from mild grade anaemia, 3 suffered from moderate anemia and 4 suffered from severe anaemia. However, no family history was observed in our study. A similar observation was made by Mohd Al Okbi et al,[21] suggesting HbS, HbF, or low haemoglobin levels did not discriminate sickle cell disease patients with sensorineural hearing loss.

CONCLUSION

Although there seems to be lower incidence of sensorineural hearing loss (SNHL) in patients with SCD, it mostly affects children of 0-2 years old. It is an obvious need to link SCD and sensorineural deafness and early diagnosis of hearing loss. Underdiagnosis or late diagnosis of hearing loss may lead to irreversible damage to the linguistic, biopsychosocial, and emotional development of patients who suffer from SCD. Persistent partial deafness distorts the speech and language acquisition during childhood. Consequentially the deaf children are academically backward and this affects their future. It seems therefore reasonable to perform a close audiological examination and follow-up by repeated audiological assessments of the patients for early rehabilitation of SNHL associated wth Sickle Cell Disease.

Disclosure of potential conflicts of interest.

No potential conflicts of interest were disclosed.

Ethical standards.

The study protocol was approved by the institutional scientific and ethical committee.

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