Journal of Advanced Medical and Dental Sciences Research

@Society of Scientific Research and Studies

Journal home page: www.jamdsr.com doi: 10.21276/jamdsr ICV 2018= 82.06

(e) ISSN Online: 2321-9599; (p) ISSN Print: 2348-6805

Original Research

Assessment of Hearing Loss in Children with Sickle Cell Anemia- A Clinical Study

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***ABSTRACT*:**
Background: Hearing deficit is described as one of the symptoms which happen because of the cochlear high sensitivity to vessel occlusion which is common in children with sickle cell anemia. The present study was conducted to assess hearing loss in children with sickle cell disease. **Materials & Methods:** The present study was conducted on 105 children with sickle cell anemia (group I) of both genders. Patients' age 5-16 years were included in the study. Equal number of controls (group II) was also considered in the study. The otolaryngology examination was carried out by ENT surgeon. The pure tone audiometry (PTA) was done by an audiometrician in a sound isolated audiometric room. **Results:** Out of 105 patients, males were 55 and females were 50. Out of 105 cases, 20 had hearing loss and in control 6 had hearing loss. The difference was significant (P< 0.05). Type of hearing loss was conductive seen in 10 in group I, 3 in group II, sensorineural seen 7 in group I and 2 in group II and mixed seen 5 in group I and 1 in group II. The difference was significant (P< 0.05). Type of hearing loss was mild seen 8 in group I, 2 in group II, moderate seen 4 in group I, 1 in group II, moderate seen 5 in group I, 2 in group II and profound 3 in group I and 1 in group II. **Conclusion:** Among children with sickle cell anemia, there was more prevalence of hearing loss as compared to control group.
Key words: Audiometry, Hearing, sickle cell anemia

Received: 13 February, 2019 Revised: 25 May 2019 Accepted: 27 May 2019

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This article may be cited as: Priyanka, Singla P. Assessment of Hearing Loss in Children with Sickle Cell Anemia- A Clinical Study. J Adv Med Dent Scie Res 2019;7(7): 90-93.

INTRODUCTION

The origin of sickle cell disease (SCD) is not known, but it is believed to have started from Africa. SCD has the ability to protect people against malaria (a deadly disease found in countries of warm weather conditions).[1] It is reported that symptoms associated with SCD crises were known by other different names in Africa way before they were identified in other continents.

Since then various studies have described different types of the disease conditions and its associated complications. The sickle-cell trait hemoglobin (Hb) S is now known to be widespread and vastly distributed, reaching its highest prevalence in parts of Africa as well as among people with origins in Equatorial Africa, Middle East, Central India, and countries bordering the Mediterranean Sea, especially Italy and Greece.[2] The disease is currently found all over Europe and in large regions of Asia besides Africa and the Americas. It is a multisystem disease, associated with episodes of acute illness and progressive organ damage and is primarily an inherited blood disorder which affects the red blood cells. However Hb SS is the commonest type of SCD throughout the world.[3]

Hearing deficit is described as one of the symptoms which happen because of the cochlear high sensitivity to vessel occlusion, causing ischemia and cochlear anoxia, because of the sickle cells which preclude blood flow to the cochlear epithelium4. The fact that the cochlea is mainly fed by one single artery, the labyrinthine artery, which can be a terminal artery, makes the inner ear very much prone to circulatory changes.[4]

Studies have reported that the deformation of the red blood cell causes the lesion to the cochlear. This prevents proper blood supply to the metabolic activity required to maintain the ionic and electrical balance of the endolymph. So the

anoxia caused to the organ of Corti would cause progressive and extensive damage to the cochlea.[5] The present study was conducted to assess hearing loss in children with sickle cell disease.

MATERIALS & METHODS

The present study was conducted in the department of Otolaryngology. It comprised of 105 children with sickle cell anemia (group I) of both genders. Patients' age 5-16 years were included in the study. Equal number of controls (group II) was also considered in the study. Exclusion criteria was patients with TB, HIV, meningitis, mumps, ear discharge and history of ear surgery, diabetes, head injury, family history of hearing loss etc. The study was approved from institutional ethical committee. All participants were informed regarding the study and written consent was obtained.

Data related to participants such as name, age, gender etc. was recorded.

All were given a semi-structured questionnaire and asked to respond accordingly. The otolaryngology examination was carried out by ENT surgeon. The pure tone audiometry (PTA) was done by an audiometrician in a sound isolated audiometric room (booth) with ambient sound less than 35 dB using a calibrated diagnostic audiometer KAMPLEX KLD 21 at frequencies 250 Hz, 500 Hz, 1 kHz, 2, 3, 6 and 8 kHz. Hearing loss was defined as present if a child has an average threshold of hearing more than 25 dB at two or more frequencies by PTA in one ear.

Results thus obtained were subjected to statistical analysis. P value less than 0.05 was considered significant.

RESULTS

Table I Distribution of patients

Total- 105		
Gender	**Males**	**Females**
Number	55	50

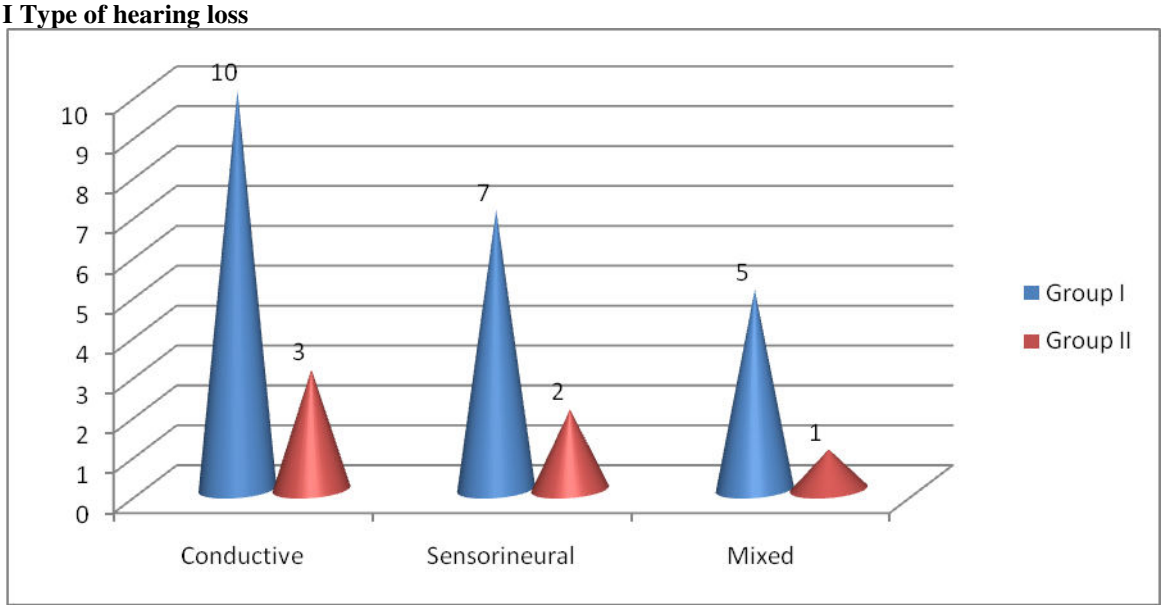
Table I shows that out of 105 patients, males were 55 and females were 50.

Table II Hearing loss in SCA patients and controls

Group I		Group II		P value
Total	**Hearing loss**	**Total**	**Hearing loss**	
105	20	105	6	0.01

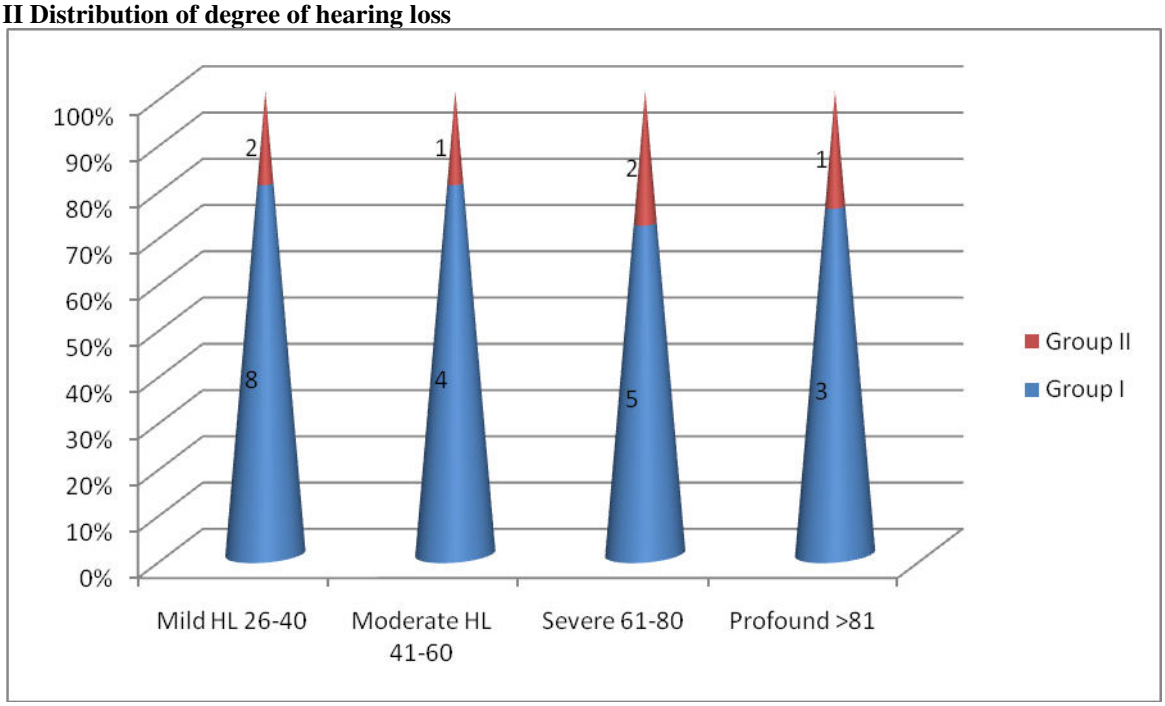
Table II shows that out of 105 cases, 20 had hearing loss and in control 6 had hearing loss. The difference was significant (P< 0.05).

Graph I Type of hearing loss



Graph I shows that type of hearing loss was conductive seen in 10 in group I, 3 in group II, sensorineural seen 7 in group I and 2 in group II and mixed seen 5 in group I and 1 in group II. The difference was significant (P< 0.05).

Graph II Distribution of degree of hearing loss



Graph II shows that type of hearing loss was mild seen 8 in group I, 2 in group II, moderate seen 4 in group I, 1 in group II, moderate seen 5 in group I, 2 in group II and profound 3 in group I and 1 in group II.

DISCUSSION

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. Although sickle cell anemia has been broadly studied in terms of population frequency and clinical manifestation, its public health issues associated with child-youth hearing loss has not received proper attention in our country.[6]

Hearing loss in SCD patients can be conductive with variable degrees of obstruction to sound transmission either in the outer ear or the middle ear, and would likely be due to susceptibility to infections in such patients.[7] Considering the delicate nature and function of the cochlea vasculature (high metabolic demand and high rate of gaseous exchanges needed for its activities), vaso-occlusion in SCD crisis can affect its functionality thereby causing sensorineural hearing loss (SNHL). Hearing loss could also be neural in nature due to abnormal transmission of nerve impulses to the acoustic nerve and central auditory process. However, the effect of vascular occlusion on hearing is normally underrated as management of SCD crisis mostly focuses on other dysfunctions of the body.[8] The present study was conducted to assess hearing loss in children with sickle cell disease.

In this study, out of 105 patients, males were 55 and females were 50. We found that out of 105 cases, 20 had hearing loss and in control 6 had hearing loss. Ndeezi G et al[9] found that between February, 2014, and March, 2015, 99 243 dried blood spots were analyzed and results were available for 97 631. The overall number of children with sickle cell trait was 12 979 (13·3%) and with disease was 716 (0·7%). Sickle cell numbers ranged from 631 (4·6%) for trait and 23 (0·2%) for disease of 13 649 in the South Western region to 1306 (19·8%) for trait and 96 (1·5%) for disease of 6581 in the East Central region. Sickle cell trait was seen in all districts. The lowest prevalence was less than 3·0% in two districts. Eight districts had prevalence greater than 20·0%, with the highest being 23·9%. Sickle cell disease was less common in children older than 12 months or who were HIV positive, which is consistent with comorbidity and early mortality.

We found that type of hearing loss was conductive seen in 10 in group I, 3 in group II, sensorineural seen 7 in group I and 2 in group II and mixed seen 5 in group I and 1 in group II. Okbi et al[10] conducted a study to study the prevalence, the pattern and associated factors of hearing loss among children with sickle cell anemia. Twenty-two of the children had hearing impairment compared to 6 controls. Prevalence of hearing loss found among children with SCA was 17% but in the control group the prevalence found was 5%. Sensorineural hearing loss (SNHL) affected 39%, conductive (CHL) 25% and mixed (MHL) 14% among cases compared to CHL 7%, SNHL 7% and MHL 7% among the controls. The hearing loss varied from mild to moderate (95%). All sickle cell children had high-

frequency hearing loss. There was a statistically significant association of hearing loss and neurologic motor deficit.

We found that type of hearing loss was mild seen 8 in group I, 2 in group II, moderate seen 4 in group I, 1 in group II, moderate seen 5 in group I, 2 in group II and profound 3 in group I and 1 in group II. It has been found that Sickle cell disease is characterized by vascular occlusive crises. Vascular occlusion can lead to compression of the auditory canal, cause damage to the hair cells of the organ of corti, stria vascularis, the basal turn of the cochlea and eventually impaired auditory function. Moreover, the sickle cell trait is extremely common in Ghana. One in every three healthy Ghanaians is either Hb AS or Hb AC. Furthermore, 2% of newborn births in (16000 annual births) present with SCD every year. However, only one study has investigated SNHL among sickle cell anemia patients in Ghana, with a prevalence rate of 29% over 25 years ago.[11]

The decrease in the levels of O2 causes morphological polymerization of the red blood cell, and it takes on an anomalous form, looking like a sickle. The elongated deformed red cells, not always can cross through small vessels, blocking them and preventing blood flow in the vicinity areas. The disease's course is variable; there are patients who have severe problems with greater frequency and others who have only sporadic health problems.[12]

CONCLUSION

Authors found that among children with sickle cell anemia, there was more prevalence of hearing loss as compared to control group.

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