SENSORINEURAL HEARING LOSS IN GHANAIANS WITH SICKLE CELL DISEASE

BY

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Summary

A study seeking to determine whether Sensorineural hearing loss occurs with similar prevalence in sickle cell haemoglobin (Hb) SS and SC disease patients was conducted on 200 subjects comprising 93 Hb SC patients, 55 Hb SS patients, and 52 Hb AA subjects serving as controls. Each subject was given haematological and pure tone audiological examinations.

Three out of 93 Hb SC patients (3.2%) showed hearing losses ranging from 30 to 40 dB but 15 out of 55 Hb SS patients (27.3%) exhibited hearing losses ranging from 30 to 60 dB. All hearing losses were in the high frequency range of 4 to 8kHz.

Further studies are required to determine why sensorineural hearing loss is prevalent in Hb SS patients but not so common in Hb SC patients.

Key Words: Audiometry; sensorineural hearing loss; sickle cell anaemia; Ghana.

Introduction

In sickle cell disease the sickled erythrocyte is rigid, mechanically fragile and has a tendency to increase the viscosity of the blood, resulting in aggregation and sludging in the microcirculation. This may lead to vaso-occlusion and haemolysis. The hallmarks of the disease are therefore recurrent intravascular haemolysis and vaso-occlusive episodes.

The incidence of the sickling gene is quite high in Ghana with a prevalence rate of 20% in the South and 10% in the North[1,2]. With this prevalence rate the expected incidence of sickle cell anaemia (SS) is around 1% in the South[3] Surveys in Ghana on the prevalence of sickle cell anaemia have provided figures ranging from 0.2% to 2.1%[4,5] sickle cell HbC (SC) disease, generally found in people of West African origin,[6] is also estimated to be found in about 1% of children born in Ghana[7]. Approximately equal proportions (48%) of sickle cell anaemia patients and patients with haemoglobin SC disease present at the Korle Bu Sickle Cell Clinic in Accra, Ghana, regularly[8]. In Ghana, therefore, the SS and SC diseases are important causes of morbidity.

Sensorineural hearing loss, a term describing any hearing impairment that originates from the footplate of stapes, is a common complication of sickle cell anaemia[9]. There is, however, hardly any information about SC patients and their levels of hearing. In Ghana, specifically, there is a dearth of information about sensorineural hearing loss in both SC and SS diseases. The purpose of this study was to find out if there are any hearing losses in our sickle cell patients.

Materials and Methods

This study was conducted on 200 subjects comprising 148 sickle cell patients and 52 controls. The patients consisted of 93 Hb SC patients (49 females, 44 males) and 55 Hb SS patients (33 females, 22 males). The average age of the Hb SC patients was 33 years (range 17 to 47 years) and that of the Hb SS patients was 24 years (range 14 to 40 years). All patients studied were regular attenders of the Sickle Cell Clinic at the Korle-Bu Teaching Hospital in Accra. The controls, made up of 23 females and 29 males had an average age of 23 years (range 12 to 42 years). They were mostly volunteers from amongst the staff (and their dependants) of the Korle-Bu Teaching Hospital.

Initial selection of subjects was done by genotyping. A questionnaire was used to

exclude all those whose sensorineural hearing losses could possibly result from other etiologies like ototoxic drugs, physical trauma, ageing, and environmental noise pollution. All subjects selected for study were healthy and had been in steady state for the preceding 3 months. None of the subjects complained of hearing impairment and there was no past history of accident and/or surgery to the middle or internal ear. Clinical otoscopic examination did not reveal any abnormality. The haemoglobin level in each subject was also determined. All results were analysed statistically using the Chi squared test.

Each subject was given a pure tone audiological examination. Hearing thresholds were determined for the octave frequencies of 250 Hz through 8000 Hz using a diagnostic audiometer MA 21 (VEB Pracitronic, Dresden, GDR). A threshold of 29 dB was taken as normal.

Results

The mean steady state Hb level was found to be 8.2 ± 0.7 g/dl (range 5.5 - 12.1 g/dl) for the 55 SS patients, 14.1 ± 0.7 g/dl for the 93 SC patients (range 11.1 - 16.0 g/dl), and 14.7 ± 0.8 g/dl (range 13.4 - 16.2 g/dl) for the 52 controls. The auditory functions of the controls were all within normal limits. However, 15 out of 55 SS patients (27.3%) exhibited hearing losses ranging from 30 to 60 dB in the high frequency range of 4000 to 8000 Hz. Only 3 out of 93 SC patients (3.2%) showed hearing losses ranging from 30 to 40 dB in the high frequency range of 4000 to 8000 Hz.

Table 1 shows the analysis of the audiological results of the affected ears from 4000 to 8000 Hz frequency range.

Table 1 further shows the distribution of the degree of hearing loss in the affected ears. A hearing loss of 30 dB was recorded from nine ears of six SS patients, three of whom showed bilateral hearing losses and the other three unilateral in two left ears and one right ear. One SC patient also showed a unilateral hearing impairment of 30 dB in the right ear. The number of the ears with 40 dB hearing loss was seven from five SS patients with two accoustically impaired bilaterally and three unilaterally, two in the left ears and one in the right; two SC patients showed bilateral hearing losses at 40 dB. All the 8 ears with 60 dB hearing loss were from SS patients whose ears were bilaterally impaired. The average degree of hearing loss in the affected ears for each experimental group has been graphically depicted in Figure 1.

Table 1

Analysis of audiological tests in the affected ears
(frequency range 4-8 kHz)

Genotype	No. of Patients	Sex (M = Male) (F = Female)	Age (Years)	Ear(s) Affected	Hearing Loss (4-8 kHz) (dB)
SC	1	M	30	Right	30
	2	M,M	27,35	Both	40
SS	1	F	21	Right	30
	2	M,F	33,24	Left	30
	3	M,M,F	31,31,29	Both	30
	1	F	26	Right	40
	2	F,F	19,37	Left	40
	2	M,F	24,22	Both	40
	4	M,M,F,F	20,26,22,32	Both	60

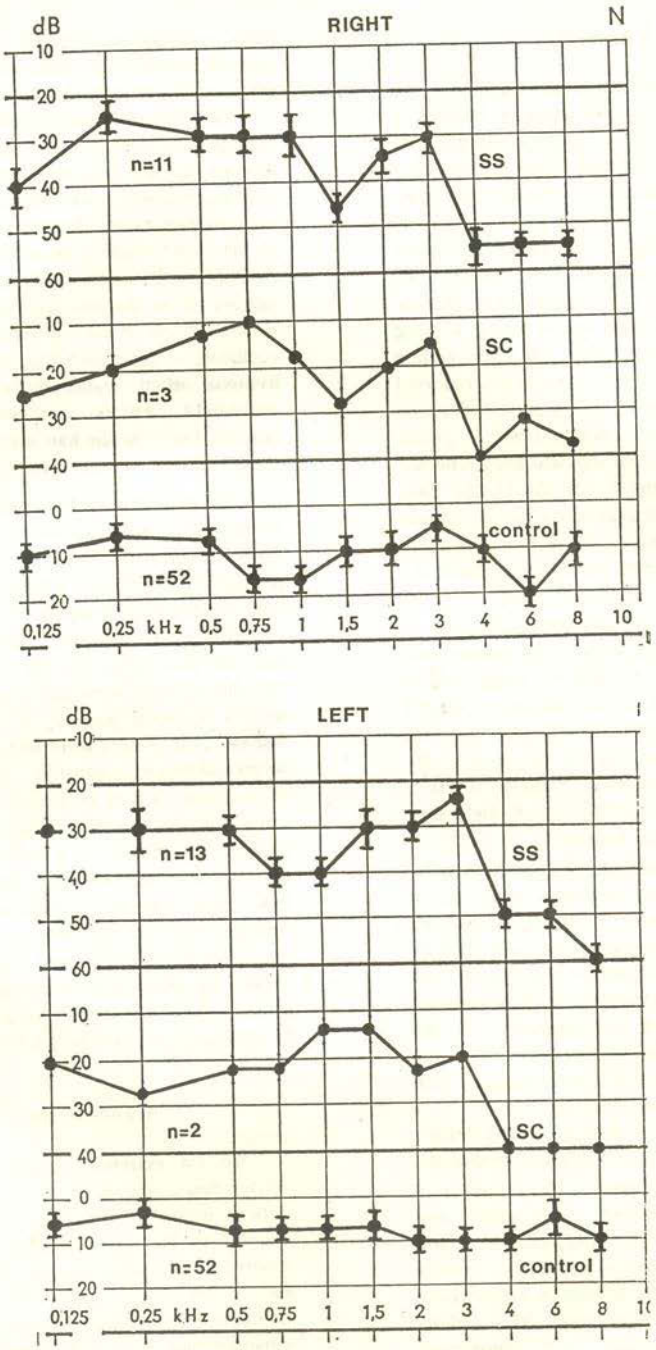

Figure 1:

Audiometric representation of the mean hearing loss in the affected ears of the different experimental groups. (n is the number of ears affected in both haemoglobin SS and haemoglobin SC patients; For controls n represents the total number of ears audiometrically examined).

Discussion

Several authors have attributed hearing loss in SS patients to sickle cell anaemia. Studies by Todd et al[9] on 83 adult Jamaican patients with sickle cell anaemia revealed a hearing loss of at least 25 dB in about 22% of the patients compared to 4% of the controls. In 1980, Friedman et al[13] described hearing loss in 12% of the children studied as opposed to a figure of 0.5% in controls. A case of a 20 year old black male who developed bilateral sensorineural hearing loss during a severe sickle cell crisis with a partial recovery after 90 days has been reported by Urban[10]. A similar case of sudden deafness related to sickle cell crisis but with a partial recovery after one year has also been reported by Orchik and Dunn[11]. In the Middle East, where the sickle cell gene is also found, Ashoor and Al-Awamy[12] have reported hearing losses ranging from 30 dB to 70 dB in 23.8% of the 42 Saudi Arabian sickle cell disease patients examined clinically and audiometrically.

Our study clearly adds to the opinion that a high frequency of auditory complications could be anticipated in patients with sickle cell anaemia. The 27.3% of our patients with sensorineural hearing loss is higher than the 23.8% and 21.7% reported in Saudi Arabia[12], and Jamaica[9] respectively although the degree of hearing loss is about the same.

The nature of the pathology underlying the functional loss is not certain. According to Morgenstein and Manace[14], compression of the auditory nerve in the internal auditory canal by the expanded bone marrow in the petrous temporal bone contributes to hearing loss; however, radiological measurements of the internal auditory canal by Sergeant et al.[15] did not show any difference between patients with and without abnormal audiograms.

Another school of thought is that ischaemia damages the cochlear. In a histopathologic study of audiometric loss in a 10 year old child with sickle cell disease, Morgenstein and Manace[14] examined the temporal bone and found extensive damage to the outer hair cells of the organ of Corti as well as degenerative changes in the stria vascularis. The authors concluded that sickling and consequent hypoxia of the inner ear produced structural changes that resulted in hearing impairment. It is also the claim of Koide et al[16] that the venous system in the cochlear has a low oxygen tension which predisposes to sickling and consequent anoxic damage. They support this claim with the observation that ligation of the vena aqueductus cochlear in experimental animals produces changes in cochlear microphonics similar to the hearing loss in SS disease.

Currently the most tenable hypothesis for the sensorineural hearing loss in SS disease appears to be impaired blood flow in the cochlear venous system during sickling. Sickle cell anaemia causes decrease in blood haemoglobin level leading to decrease in oxygen delivery to the auditory organ; this is further aggravated by the episodic vaso-occlusion that occurs because of the low oxygen tension in the cochlear; consequently, tissue and nerve hypoxia when sustained beyond a certain threshhold causes permanent neurologic changes either in the hair cells of the organ of Corti or in the auditory nerve.

Although this study does not establish the mechanisms for the sensorineural hearing loss in sickle cell anaemia, it confirms that a significant proportion of patients with sickle cell anaemia have sensorineural deafness.

The same, however, cannot be said of patients with sickle cell Hb C disease which, like the controls, were not anaemic. Although sudden deafness had been reported in SC disease[17], our results seem to indicate that sensorineural hearing loss is not a major problem in SC patients. It may be argued that the relative rarity of sensorineural hearing loss in SC patients may be due to a reduced tendency of the red cells in SC patients to sickle leading to a lower morbidity in Hb SC disease.[3,18] The damage caused to the cochlear tissues through vaso-occlusion is therefore minimised. Further studies are required to determine why sensorineural hearing loss is more of a problem in Hb SS patients than in Hb SC patients.

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