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Sensorineural hearing loss in adults with sickle cell anaemia

PA Onakoya, OGB. Nwaorgu and WA. Shokunbi=
Department of Otorhinolaryngology, =Department of Haematology, University of Ibadan, Ibadan, Nigeria.

Summary
Sensorineural hearing loss (SNHL) is one of the known complications of sickle cell disease (SCD). However, there is paucity of information on SNHL as a complication of SCD, especially in sickle cell anaemia (SCA) in our environment, hence this study. This was a prospective study of pure tone audiological assessment of 167 adult SCA patients in stable condition attending the adult Sickle Cell Clinic and 100 apparently healthy Haemoglobin AA adults as control in the University College Hospital, Ibadan. Their ages ranged from 15 to 56 years for SCA and 15 to 65 years for the controls, with a mean age of 24.2 (± 8.2) and 28.7 (± 11.9) years respectively. There were 94 females (56.3%) and 73 males (43.7%), fifty - two females (52%) and forty - eight males (48%), with a M : F ratio of 1 : 1.3 and 1:1.1 for SCA and controls respectively. Sensorineural hearing loss (SNHL) was observed in a total of 178 ears in 110 SCA patients and 68 ears in 47 controls with a prevalence of 66% and 47%, respectively. Sixty-eight patients (62%) and twenty-one controls (44.7%) had bilateral impairment, although only 18 SCA patients (11%) perceived hearing impairment. High frequency loss (4000 – 8000Hz) was commonly affected in both the SCA patients and controls as compared to other frequency ranges. Low frequency range was involved in ten ears (9%) especially the right ear of some SCA patients..Decibel hearing level (dBHL) loss was in the mild range (26 – 40 dBHL) in 103 (58%) and 53 (78%) ears in the SCA and controls, respectively. Five patients had severe and profound dBHL loss. The range of dBHL loss was 26 – 43 dBHL especially in the high frequency range bilaterally for both the SCA and controls. Mean binaural hearing of 13 dBHL was recorded in both the SCA and controls for each octave frequency bilaterally in those with normal hearing while 26 and 23 dBHL were for those with impaired hearing respectively. Also, the mean dBHL for both ears was observed to be progressively worse with increasing age groupings, more especially in SCA patients. There was no significant correlation between the severity of hearing loss and the frequency of vaso-occlusive crisis. It is hoped that this study would have increased the awareness that SNHL is a common complication of SCA in our patients. There is thus the need for periodic evaluation of the auditory function of SCA patients in our environment.

Keywords: *Sensorineural, Hearing loss, sickle cell anaemia.*

Résumé
La perte d'ouïe sensorineural (POSN) est une complications de la maladie drépanocytose (MD) connue. Pourtant, il y a une disette d'information sur POSN comme une complication de MD en particulier à ce qui concerne l'anémie à hématies falciformes (AHF) dans notre milieu ainsi l'étude cette étude est une étude en prospective d'une évaluation purement endiologique des 167 adultes malades de AHF dans une conditions stable qui fréquentent la clinique drépanocytose et d'un groupe de contrôlede 100 personnes qui sont evidemment en bonne HaemoglobinAA dans le centre hospitalier universitaire. Leurs âges varient entre 15 à 56 ans pour les AHP et le groupe de contrôle avec la moyenne âge de 24,2 (±8,2) et 28,7 (±11,9) ans respectivement. Il y avait 94 femmes (56,3%) et 73 hommes (43,7%), cinquante deux femmes (52,3%) et 73 hommes (43,7%), cinquante deux femmes (52%) et quarante-huit hommes (48%) avec une proportion d'homme et femme de 1 :1,3 et 1 :1,1 pour AHP et aussi pour le groupe de contrôle. La perte d'ouïe sensorineural dans 178 oreilles de 100 malades AHP et 68 oreilles chez 47 de personne de groupe de contrôle avec une fréquence de 66% et 47% respectivement. Soixante huit malades (62%) et vingt un de groupe de contrôle (44,7%) avaient une déficience bilatérale. Bien que surtout 18 malades AHP(11%) ont remarqué une déficience auditive. Une perte de haute fréquence (4000-8000Hz) était plus affecté chez les malades de AHP et les contrôles par comparaison à d'autre proportion de fréquence. Une proportion inférieur de fréquence était noté dans dix oreilles (9%) en particulier dans l'oreille droit de quelques malades AHP du nivaux de l'ouïe décibel (NodB) était a portée légère (26-40 NodB) en 103 (58%) et 53 (78%) oreilles chez les AHP et les contrôle respectivement. Cinq malades ont eu une perte NodB sévère et profond. La proportion de NodB était 26-43 surtout dans le cas de proportion de fréquence élevés pour les AHP et les contrôles. La moyenne d'ouïe binaura de 13 NodB était noté chez les AHP et les contrôles pour chaque fréquence octave bilatéralement chez ceux d'ouïe normal tandis que 26 et 23 NodB sont pour ceux avec l'ouïe affaiblie respectivement. Ensuite, on a noté que la moyenne NodB pour les deux oreilles se détériorent avec l'augmentation progressive des groupes d'âge, en particulier chez les malades AHP. Il n'y avait pas une corrélation entre la gravité de perte d'ouïe et la fréquence de crise vaso-occlusive. Nous espérons que cette étude aurait augmenté la conscience que le NodB est une complication commune de AHP chez nos malades. Il y a donc la nécessité d'une évaluation périodique de la fonction auditoire des malades AHP dans nos milieu.

Correspondence: Dr. P.A. Onakoya , Department of Otorhinolaryngology. University College Hospital, Ibadan, Nigeria.

Introduction

Sickle cell disease (SCD) is a common haematological disorder of clinical importance. It is common especially in people of African descent in the endemic malarial zones of Central and West African sub-regions. It is also worthy of note that many of the sickle cell anaemia (SCA) patients are now living longer into adulthood due to improvedment management.

Generally, SCD runs an extremely variable clinical course, characterized by haemolytic anaemia interspersed by severe exacerbation of haemolysis (haemolytic crisis), vaso-occlusive disorders (e.g., bone pain and abdominal crises, priapism), sequestration crisis and acute chest syndrome, with complications that are usually due to repeated episodes of vascular occlusion. In the central nervous system, increased sensitivity of tissues or organs to hypoxia leads to infarction following vascular occlusion[1].

The involvement of the ear in these processes as seen in SCD patients usually leads to hearing impairment especially sensorineural hearing loss (SNHL) as was first reported by Diggs[1]. Morgestein and Manace[2] observed that histological examination of temporal bone of a ten-year-old child with audiometric

loss was consistent with changes seen in the stria vascularis and the hair cells of the organ of Corti, which may be due to hypoxia.

In this environment, there is limited emphasis placed on the hearing of adult SCD patients especially SCA in the steady state. Hearing loss can result in considerable disability both for learning and communication; there is the need for early detection and prevention in this group of patients. The aim of this study was to determine the pattern of hearing loss and to identify any correlation between frequencies of vaso-occlusive crisis and hearing impairment among the adult SCA patients, especially in steady state as seen in the University College Hospital, Ibadan, Nigeria.

Patients and methods

This prospective study was conducted among 167 adult SCA patients in steady state attending the adult sickle cell clinic of the Haematological Department, University College Hospital, Ibadan. One hundred apparently healthy Haemoglobin A (AA) adults who did not have any referable complaints to the ear were randomly selected from different outpatient clinics, among the hospital workers and students to serve as control. The standard requirements of the Ethical Committee of the Hospital were met. Informed consent was obtained from each patient and control and their ages ranged from 15 to 56 and 15 to 65 years, respectively.

Relevant histories such as age of onset of features suggestive of SCA, frequency of crisis per year and any perception of hearing loss and duration were obtained at the time of first review using a structured questionnaire. Detailed clinical and otological examinations were done for the SCA patients and the control. Any observed wax in the external auditory canal was removed and thereafter the patient was re-assessed otologically.

Pure tone audiometry in a sound proof booth using an audiometer (Ampivlox 2150) which has been calibrated to ISO standard was done at 0 and 3 months interval for the SCA patients and once for the controls, with an ambient noise level of 25 decibel hearing level (dBHL) outside the booth (measured using sound level indicator CS-15E).

The mean hearing threshold for each octave frequency from 125 to 8000 Hertz (Hz) was then determined and the degree of hearing loss for each subject was determined according to the WHO standard classification[3].

Excluded from this study were patients who had:

1. History of blood transfusion 6 weeks prior to evaluation, which may suggest recent sickle cell crises as only patients in steady state were
2. Congenital ear anomalies.
3. recruited for the study.Acquired ear lesions such as infection, inflammation, tumours, sequelae of otitis media, previous ear surgery and ototoxic drug usage.
4. History of systemic diseases such as cerebrovascular accidents (cva), diabetes mellitus, tuberculosis, or any acute illness at the time of review.
5. Head injury or trauma to the ear and familial deafness.

The data were analysed using the SPSS 7.5 package for the appropriate statistical analysis and cross-tabulation for the correlation. All the results were presented in tabular forms.

Results

A total of 167 SCA (SS) patients and Haemoglobin A adults mean age of 24.2 (± 8.2) and 28.7 (± 11.9) years, respectively. There were 94 females (56.3%) and 73 males (43.7%), fifty-two females (52%) and forty eight males (48%), with a M : F ratio of 1 : 1.3 and 1:1.1, respectively.

Table 1: Frequency of vaso-oclussive criss

Crisis per year	Sca = 167	(%)
0 - 5	134	(80.2)
6 - 10	27	(16.2)
≥ 11	2	(1.2)
Not know	4	(2.4)

The mean number of bone pain crises per year was 4.1 ± 3.4 (range 1 – 25) and 134 patients (80.2%) were observed to have between 0 and 5 crises per year (Table 1). The mean age at first presentation of symptoms of SCA was 3.7 ± 3.2 years (range 4 months – 24 years).

Table 2: Ditribution of ears in the affected patients

EAR	Sca = 110 (%)	AA = 100(%)
RIGHT	22 (20)	20(42.5)
LEFT	20 (18)	6 (12.8)
BOTH	68 (62)	21 (44.7)

No of Ears = 178 and 68 for SCA & control (AA) with (63% and 24% of total num. of ears respectively.

Only 18 patients (11%) could perceive hearing impairment out of which 10 could give the duration of their impairment. The mean duration of the impairment was 39.6 ± 32.0 months (range 12 – 120 months).

A total of 178 ears in 110 SCA patients and 68 ears in 47 controls were judged to have hearing impairment thus giving a prevalence of 66% and 47% respectively. Sixty-eight patients (62%) and twenty-one controls (44.7%) had bilateral impairment (Table II). High frequency loss (4000 – 8000Hz) was commonly affected in both the SCA patients and controls as compared to other frequency ranges (Table 3). It was also observed that the low frequency (125 – 250Hz) was involved in 10 ears (5.6%) especially in the right ear of some of the SCA patients.

Table 3: Distribution of hearing impairment at various frequencies in the affected ears

Frequency (Hz)	Level of Hz.	Sca = 178(%)	AA=100(%)
High	4000-8000	82 (46)	37 (54.4)
Middle	500-2000	2 (1)	3 (4.4)
Low	125-250	10 (6)	3(4.4)
All freq.	125-8000	32(18)	17(25.0)

Decibel hearing level (dBHL) loss was observed in the mild range (26 – 40 dBHL) in 103 (58%) and 53 (78%) ears in the SCA and controls respectively. Five patients had severe and profound dBHL loss (Table 4). The range of dBHL loss in this study was 26 – 43 dBHL especially in the high frequency range bilaterally for both the SCA and controls.

Mean binaural hearing of 13 dBHL was recorded in both the SCA and controls for each octave frequency bilaterally in those with normal hearing while 26 and 23 dBHL were for those with impaired hearing respectively. The mean dBHL was ob-

served to be progressively worse in both ears with increasing age groupings in both SCA patients and controls, but more in the former (Table 5). There was no significant correlation between the frequency of vaso-occlusive crisis and impairment of hearing (Table 6).

Table 4 : Distribution of decibel hearing level loss among the affected ears

dBHL Loss	Range	Sca=178(%)	AA=68(%)
Mild	26-40	103 (58)	53 (78.0)
Moderate	41-60	16 (9)	5(7.3)
Severe	61-80	4 (2)	0(0.0)
Profound	≥ 81	1 (1)	0 (0.0)
Other combinations		54 (30)	10 (14.7)

Table 5: Mean decibel hearing level at 8KHz for different age groups (SCA patients and controls (AA) respectively

Age group (Year)	Number of Patients	Mean decibel Hearing level Left ear	Right ear	Number of Patients	Mean decibel Hearing level Left ear	Right ear
15-24	103	23.2	25.5	46	16.6	18.4
25-34	47	36.7	35.4	28	20.0	21.8
35-44	11	40.9	43.6	14	31.1	30.4
45-54	5	53.0	49.0	7	29.3	37.9
55-65	1	60.0	55.0	5	50.0	49.0
Total	**167**	29.3	30.4	**100**	22.2	23.9

Table 6: Correlation between frequency of vaso-oclussive crisis and impairmen of hearing in sca patients

Frequency of vaso-occlusive	Normal Hearing	Impaired Hearing	Total	Normal Hearing	Impaired Hearing	Total
0 - 5	73	62	135	70	65	135
6 - 10	9	18	27	9	18	27
11	2	3	5	2	3	5
Total	84	83	167	81	86	167
x^2		0.153			0.136	
df		2			2	
P value		0.048			0.081	

Discussion

The importance of SCD as a major cause of SNHL needs to be emphasised especially as there is paucity of information on the prevalence of SNHL in SCA patients in addition. There is a need to increase the awareness of those involved in the management of these patients in our environment.

Even the awareness of SNHL in the patients themselves is very low as was observed in this study in which only 11% could volunteer such history. The reason for this could be due to the non-acceptance and/or ignorance of hearing impairment until attention is drawn to it during interactions with others or close associates. It may also be due to greater preoccupation with other serious multisystemic complications of SCA that they often experience.

It is noteworthy that many SCA patients are now living longer due to improved education and health facilities available. Varying degrees of SNHL have been observed in the literature, with prevalence ranging from 12 to 27% [2,4-6]. In the study similar to ours among the adult age group (15 years and above) by Tsibulevskaya et al[7], a prevalence of 44.8% was observed while it was 66% in ours.

Some authors have noted that the adult age group is at an increase risk of developing severe SNHL[4,7-9]. This was equally observed in our study in which the SNHL progressively got worse with age when compared with the controls, not minding the fact that they are likely to have been exposed to the same environmental hazards like noise in our environment. However, Friedman et al [4] did not find an upward trend in the incidence of SNHL with age, although they opined that this might be due to the small population size of their study.

There was no correlation between the severity of SNHL and the frequency of vaso-occlusive crisis. This may be due to the fact that majority of these patients could have had most of the damage done during the early stage of their ailment, especially in childhood due to greater or higher frequency of attacks and various forms of crisis.

The theory of hypoxia in the cochlea predisposing to sickling and infarction with resultant SNHL was proposed by Koide et al[1]. Kimura and Pelman[10] also observed that venous rather than arterial obstruction causes progressive damage to the cochlea, especially in the basal region over a time period. Various workers have put forward theories as to the sites of damage, whether cochlear or retrocochlear[6,9,11,12]. However, tests such as the middle ear compliance, acoustic reflexes and other relevant tests to identify those due to either cochlea or retrocochlear damage were not done in this study.

SNHL in SCA has been found to be commonly bilateral[5,13]. This was confirmed in 62% of the patients in this study. This shows an equal exposure of both ears to the same pathological changes due to cochlea venous occlusion over a period of time, without any predilection for lateralisation.

SNHL commonly occurs in the high frequency range (2000 – 8000Hz) with varying degrees of dBHL loss from mild to profound (total). It was however noted in this study that varying degrees of dBHL loss ranging from 45 to 70dBHL occurred at the high frequency range (4000 – 8000Hz). However, of the total number of ears, 46% had high frequency loss while 58% were in the mild dBHL loss range. The range of the dBHL loss was 26 – 43dBHL at 4000 – 8000Hz, which is similar to the findings in some earlier studies[4,7].

It was also observed that the low frequency range (125 – 250Hz) was affected in some patients. This may be in support of diffuse damage to the cochlea organs rather than basal turn only. The average binaural dBHL for those within the normal range was 13dBHL, while those with SNHL were 26dBHL especially in the high frequency range, similar to earlier reports on SCD patients with SNHL. However, this was slightly different from those recorded by Tsibulevskaya[7] in which those with normal hearing and those with SNHL were 15.2dBHL and 20.1dBHL respectively.

In conclusion, it is hoped that this study will create awareness that SNHL is a common complication of SCA in our environment. There is thus the need for periodic assessment of the auditory function of these patients especially from pre-school age period, because of the effect of SNHL on their progress in school as well as their social interactions. There is also the need for further research into the pathological factors that may be responsible for the high increase in the prevalence of SNHL among the SCA and more especially the SCD patients in our environment. This will serve as a guide towards a more focused approach to their comprehensive management.

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