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Audiometric Findings in Adolescent Sickle Anemia Patients in Port Harcourt

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Abstract

Background: Sickle Cell disease (SCD) is a hereditary disease of the erythrocyte in which the propensity to form rigid sickle morphology is increased. Hearing loss is one of the central nervous system complications of sickle cell disease.

Aim: The aim of this study was to evaluate the proportion of adolescent SCD patients with hearing loss in Port Harcourt,

Objective: To determine the severity of hearing loss among SCD patients and to determine socio -demographic factors related to SCD patients with hearing loss.

Methodology: This is a comparative cross sectional study of 60 adolescent SCD patients and 60 age and gender matched HbAA controls. Otoscopy, tympanometry and pure tone audiometry were done on these patients.

Results: Among 60 adolescent SCD patients aged 10-19 years, 4(6.7%) had sensorineural hearing loss whereas 1(1.7%) of age and gender matched HbAA controls had sensorineural hearing loss. All cases of hearing loss were mild in severity. The mean hearing threshold was higher in the SCD patients than control for most frequencies tested.

Conclusion: Most of the adolescent sickle cell patients had normal hearing. Neither age nor gender was found to have correlation with hearing loss among the SCD patients. Mean hearing threshold was higher for the sickle cell patients than control.

Keywords: Pure tone; Adolescents; Sickle Cell; Hearing loss; Anemia

Abbreviations: SCD: Sickle Cell Disease; Hb: Haemoglobin; RBC: Red Blood Cells; CNS: Central Nervous System

Introduction

Sickle cell disease (SCD) is an inherited chronic haemolytic anemia whose clinical manifestations arise from the tendency of the haemoglobin (Hb) to polymerize and deform the erythrocyte into characteristic sickle shape [1]. Sickling is due to single nucleotide change in the β- globin chain leading to substitution of valine for glutamic acid at position 6. In SCD, an individual inherits two abnormal haemoglobin (Hb) genes, at least one being HbS [1,2]. HbSS is most common form and is known as sickle cell anemia [2]. Other less common forms are HbSC, HbSβ-thalassaemia, HbD Punjab and HbO arab [3].

Globally it is estimated that 20-25 million people are living with the homozygous SCD [4]. It is estimated that 240,000 are born with SCD in sub-Saharan Africa annually [4]. Nigeria has the greatest number of sickle cell disease patients in the world [5]. About 2% of all newborns in Nigeria are said to be born with the disease [6]. The normal red blood cells (RBC) are pliable and oval in shape, which contributes to their smooth flow through the vessels [1,2]. In contrast, when RBCs in patients with SCD are exposed to any stress like dehydration, deoxygenation, cold temperature or infections, they become hard and sickle in shape [1,3]. This change significantly hinders their flow through small vessels, which in turn leads to obstruction, ischemia and end organ hypoxia. This manifests clinically as Vaso occlusive crisis which may affect different systems of the body [2].

One of the vulnerable systems of the body is the central nervous system (CNS) [4]. This is due to poor tolerance to low oxygen tension by CNS. Peripheral auditory dysfunction is one

of the common CNS complications seen in SCD [8-10]. Hearing is the ability to perceive sounds by detecting vibrations and changes in the pressure of the surrounding medium through time [11]. Normal hearing is important for language development, interpersonal communication, acquisition of new knowledge and responsiveness to environmental sounds. Hearing impairment refers to total or partial inability to hear sound. Over 5% of the world population or 466 million people have disabled hearing loss and most of them live in third world countries [12]. About 8.5 million Nigerians have disabling hearing loss, 3.5 million of them being children aged between 0- 15 years [13].

Irrespective of the age at which hearing loss develops, disabling hearing loss has devastating consequences for psychosocial wellbeing, quality of life and economic independence [14]. Several causes of hearing loss have been reported, SCD is one of them. The prevalence of hearing loss among SCD patient in the United States of America (USA) and Jamaica is 12% and 22% respectively [15,16]. In Asia, the Prevalence of hearing loss among SCD patients has been found to be 37% in Oman and 19% in Saudi Arabia [17,18]. In Africa, the prevalence is 36.5% in Kenya and 60% in Ghana [19,20]. In Nigeria, two studies among children with SCD gave prevalence rate 3.8% and 13.4% respectively [9,21]. While the prevalence among adult SCD patients ranges between 4.3-66% [8,22]. So much of human endeavor relies on sound to convey meaning and losing it inevitably omits the adolescent from a wider realm of understanding [11].

Occurrence of hearing loss in the adolescent SCD patient will have untoward effect in the progression to adulthood. Inappropriate response, failing to respond at all or misunderstanding of spoken words seen in people with hearing loss can lead to social withdrawal and thus erode their stability and emotional wellbeing [11]. When SCD is complicated by hearing loss, there will be further decline in academic performance and survival skill acquisition [23]. Bees et al. demonstrated that children with mild hearing impairment performed poorer than aged match normal controls in series functional test [23,24]. Hence early diagnosis and treatment of hearing loss may serve as major boost to learning and skill acquisition.

Material and Method

The study is a comparative cross sectional study among adolescent sickle cell patients attending Outpatient Sickle cell in two tertiary hospitals in Port Harcourt. Age and gender matched normal volunteer controls were recruited from a Secondary School in Port Harcourt and first year undergraduates of University of Port Harcourt aged between 10-19 years. Sickle Cell Disease patients with pre-existing otological diseases and Aden tonsillar diseases were excluded from the study. The patients recruited for this study had otoscopy, pure tone audiometry and tympanometry done on them. The sample size was calculated using formular n = $(Z\alpha + Z\beta)^2$ P(1-P)/ $(P1-P2)^2$. Random sampling technique was used to recruit patients. A list of all eligible SCD patients booked for the day was obtained per clinic day.

Random numbers were then generated electronically using winpepi software. This random numbers were used to select the corresponding patients from the list. Questionnaires were administered to patients who gave consent. Information that was sought included patient's age, gender, level of education, history of hearing loss, family history of hearing loss, postnatal history. The controls' genotypes were confirmed by haemoglobin electrophoresis test. Each of the participants had otoscopy, tympanometry and pure tone audiometry. Tympanometry was done using tympanometer (battery powered autotymp Tm 262 Welch Allyn serial number 988964 USA). Pure tone audiometry was carried out in a sound proof booth by the researcher after adequate training. Diagnostic audiometer 229 by interacoustics serial number 160252 with well-fitting earphones was used.

Measurements of pure tone frequencies (Figure 1) 125, 250, 500, 1000, 2000, 4000 and 8000 Hz on each ear were carried out. Bone conduction was done with appropriate bone vibrator placed on the mastoid on each ear. Pure tone averages were calculated for (500, 1000, 2000 and 4000Hz). Results were used to grade hearing threshold into normal, mild, moderate, severe and profound hearing according to WHO classification [25]. Data analysis were done using SPSS version 20. The pure tone findings were analyzed for the presence, type and severity of hearing loss. Categorical variables were compared using chi-square and fisher's exact as appropriate. The level of significance was set at a p-value of less than 0.05.

Result

A total of 60 SCD patients, aged 10-19 years with mean age 14.85 ±2.944 years were recruited into the study. They were matched with 60 HbAA healthy controls, aged between 10-19years. The median age in this study was 15 years (Table 1,2 and 3).

Discussion

In this study 6.7% of the SCD patients and 1.7% of the controls had hearing loss, this was however not statistically significant. The majority (93.3%) of the adolescent sickle cell disease patient had normal hearing. This means that hearing loss is not a common complication of sickle cell disease in Port Harcourt. This outcome is similar to what was observed by Mgbor and Emordi in Enugu [9]. Most (86.6%) of their adolescent sickle cell patients had normal hearing while only 13.4% had hearing loss. Their study was among 56 homozygous (HbSS) sickle cell disease patients aged between 6-19 years. The similarity in demographics (same age group) and haemoglobin genotype (HbSS) may be responsible for the similarity in complication rate. However higher proportion

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hearing loss was observed by Onakoya et al. in Ibadan Nigeria [22]. They studied 167 adult SCD patients aged 15 to 56 years and found hearing loss in 66% of them. This high proportion of hearing loss may be due to the adult population in which their research was carried out.

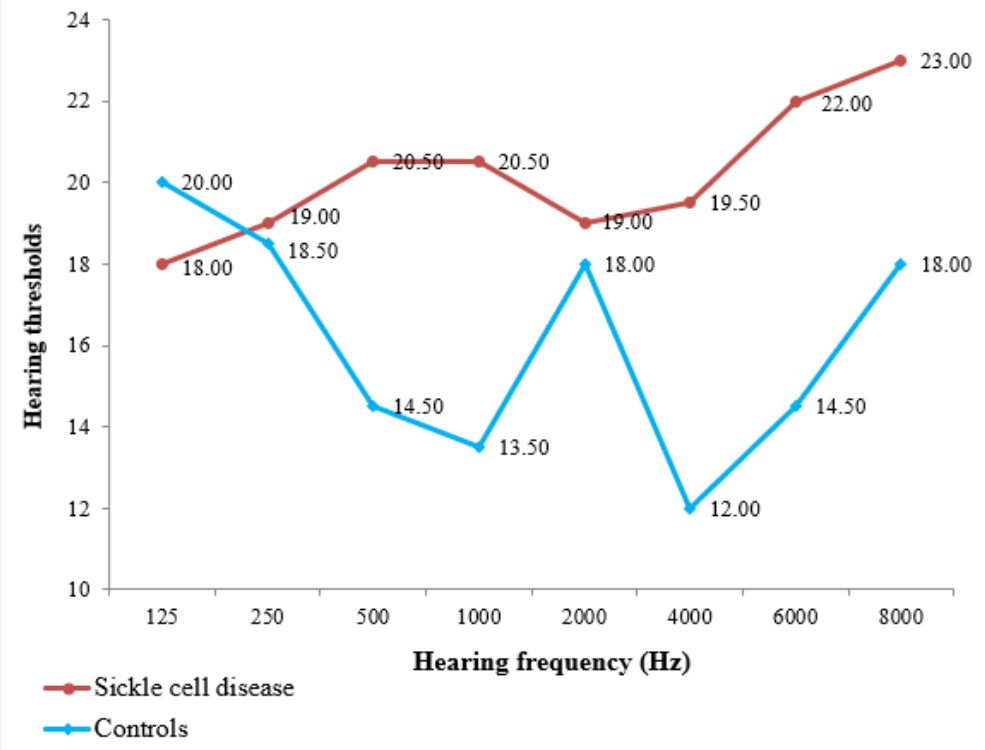

Figure 1: Measurements of pure tone frequencies.

Table 1: Age Distribution of Patients and Controls.

Age Category	Sickle Cell Disease n (%)	Controls n (%)	Total n (%)
10-14 years	26 (43.3)	22(36.7)	48(40.0)
15- 19 years	34 (56.7)	38(63.3)	72(60.0)
Total	60 (100)	60 (100)	120(100)

$\chi 2$ =0.556; p-value =0.456.

Table 2: Gender Distribution of SCD Patients and Controls.

	Sickle Cell Disease n (%)	Controls n (%)	Total n (%)
Male	28(46.7)	31(51.7)	59(49.2)
Female	32 (53.3)	29(48.3)	61 (50.8)
Total	60(100)	60(100)	120(100)

$\chi 2$=1.166; p-value=0.28.

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Table 3: Proportion of Hearing Loss in Sickle Cell Disease Patients and Controls.

	Sickle Cell Disease n (%)	Control n (%)	Total n (%)
Present	4 (6.7)	1 (1.7)	5 (4.2)
Absent	56 (93.3)	59 (98.3)	115 (95.8)
Total	60 (100.0)	60 (100.0)	120 (100.0)

Fisher's exact p-value=0.207.

Among the adult population the chance of hearing loss occurring from other causes like occupational noise exposure, ototoxicity, hypertension, diabetic mellitus and presbycusis was higher than adolescent [26-28]. In Ilorin, north central Nigeria, Alabi et al. [21] gave a lower proportion of hearing loss among the sickle cell disease patients they studied [21]. They studied 80 SCD children with mean age 9.4 years and found sensorineural hearing loss in 3.8% [21]. They also reported conductive hearing loss from otitis media with effusion in 27.5% of the SCD patients. The proportion of sensorineural hearing loss was comparable to the index study possibly because of the young age (4-19) in which their study was carried out. In Kenya, Tsibulevskaya et al. [20] documented hearing loss in 40% of their SCD patients [20]. They studied 62 SCD patients aged between 7-30 years. The predominant sickle cell haplotype in Kenya is CAR (this runs the worst disease course). The overall risk of organ failure caused by obliterative sickle cell vasculopathy is threefold in patients with CAR haplotype than others.

In Greece Koussi et al. [29] studied 24 SCD children with mean age 12.5 years [29]. Only one of their patients had hearing loss. Most of the SCD patients in their study were Sβ- thal (disease course is said to be mild). That may have accounted for the low rate of hearing in their study. Majority (98.3%) of the adolescent HbAA controls in this study had normal hearing. This shows a marked improvement compared to previous work done by Onotai et al. [30] in the same environment [30]. They studied hearing loss among primary school pupils in Port Harcourt and found hearing loss in 29.4%. The reduction in proportion of hearing loss among adolescents in this study may be due to the inclusion criteria. Only adolescents with HbAA were included while other haemoglobin genotypes were excluded.

In this study, the mean hearing threshold value for SCD patients were consistently higher than the controls for most frequencies measured. Mean threshold value for the SCD patients was significantly higher than controls at 500 and 1000Hz in the right ear. The fact that mean hearing threshold in SCD patients was higher than controls in most frequency may suggest that hearing impairment remains a problem to SCD patients in our environment. Perhaps at a later age their hearing threshold may deteriorate. All four patients who had hearing loss in this study were mild in severity. No severe or profound hearing impairment was seen. This agrees with findings of earlier researchers on severity of hearing loss among SCD patients [8,15]. Onakoya et al. found mild hearing loss in most of their patients while Aderibigbe et al. reported mild hearing loss in all cases of hearing impairment. However, Mace et al. in 2009 reported profound sensorineural hearing loss in a 7 year old sickle cell anemia patient following acute Vaso- occlusive crisis.

All four cases of hearing impairment were sensorineural in nature. This agrees with the findings of most studies. Burch - Sim and Matlock reported sensorineural hearing loss of cochlear origin. They carried out pure tone audiometry and otoacoustic emission on 183 SCD patients. The current study could not state if the sensorineural hearing loss observed is of cochlear or central origin because otoacoustic emission and auditory brainstem evoked response audiometry were not used in the study. Alabi et al. found both conductive and sensorineural hearing loss. Conductive hearing from otitis media with effusion was seen in 27.5% of their patients. The index study did not record any case of conductive hearing loss from otitis media, possibly because patients with abnormal tympanic membrane and tympanogram were excluded from the study.

Three out of the four cases of hearing loss in our study were aged between 15-19years (oldest age group). There was no statistical significance between the proportion of hearing loss and increasing age. However, Onakoya et al. and Aderibigbe et al. found statistically significant relationship between occurrence of hearing loss and increasing age [8,22]. In this study, most of the SCD with hearing loss were females, although, this was not statistically significant. This agrees with the work by Aderibigbe et al. Mgbor et al. and Al-Okbi et al. [8,17]. They found hearing loss to be commoner in female SCD patients [8]. It is possible that chronic anemic state of sickle cell disease and monthly menstrual loss in females may predispose to worse cochlear blood supply during Vaso occlusive crisis.

In conclusion, most of the adolescent sickle cell patients had normal hearing. Neither age nor gender was found to have correlation with hearing loss among the SCD patients. Mean hearing threshold was higher for the sickle cell patients than control. This study used pure tone audiometry and found only sensorineural hearing loss. We recommend newer modalities

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like otoacoustic emission and auditory brainstem response audiometry (ABR) to test hearing among adolescent sickle cell patients. These modalities are more sensitive and give the exact location of the pathology.

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