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Otological burdens of Nigerian children with sickle cell disease

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ABSTRACT

Introduction/Objective: Sickle cell disease (SCD) is associated with episodic illnesses, multi-systemic affectations and end-organs damages. Otolaryngological related complications are not unexpected. Studies on the overall Otolaryngological pathologies in children with SCD relative to their non-SCD counterparts are scanty in Nigeria. We hypothesized that children with SCD are likely to have more otological burdens than their non-SCD counterparts. Thus, we embarked on this study to describe and compare the overall ear diseases burdens seen in children with sickle cell disease relative to their non-SCD counterparts.

Methodology: A cross-sectional study of otologic diseases among children with SCD and their non-SCD counterparts attending the paediatrics and otolaryngological clinics of a Nigerian tertiary institution was conducted.

Results: Overall, 80 (47.62%) of the 168 ears of SCD patients compared to 37 (22.02%) of the 168 ears of their non-SCD counterparts were affected by diseases ($p < 0.0001$). The diseases were Wax, Otitis Media with Effusion, Suppurative Otitis Media, Otosclerosis and Sensorineural Hearing Loss (SNHL). There was a significant difference in the prevalence of SNHL and solitary otosclerosis between the SCD patients and their non-SCD counterparts ($P < 0.05$) respectively. Both the Haemoglobin concentration and HbF did not discriminate between the SCD participants with or without SNHL ($P > 0.05$).

Conclusion: This study showed that otological burdens are more prevalent in children with SCD than the non-SCD population. The microbiological peculiarity of suppurative otitis media (SOM) among participants stresses the need for concerted efforts at preventing SOM in SCD children. There is need for special Otolaryngological care for SCD children.

1. Introduction

Sickle cell disease (SCD) is a common genetic disorder that is predominantly found in Africa. It is now being increasingly recognised the world over and Nigeria is the country with the highest burden of the disease in the world [1–5]. Although, the origin is unknown, it is known to affect mainly the mixed race population [6]. The structural abnormality in SCD leads to sickling of the red blood cells at low oxygen tension. This results in clogging at microvascular level and subsequent vaso-occlusion, hypoxia and other clinico-pathologic manifestations of the disease [1,7–9]. Chronic haemolysis and anaemia, increased susceptibility to infections and recurrent episodes of vaso-occlusions manifesting as painful crises and other untoward events are cardinal manifestations in SCD [1]. Also associated with the disease are otologic manifestations and various forms of sensorineural hearing loss [2–12]. The hearing loss could be unilateral or bilateral [8,13], insidious or sudden [8,13], reversible or irreversible [14] and of varying degrees [8,13,14]. Although, most of the sensorineural hearing loss are said to be mild, there are reports of patients who developed severe to profound sensorineural hearing loss in whom cochlear implant is required to restore hearing [3,15]. Irrespective of the occlusive vascular involvement, audiometric assessment is a sensitive indicator of any sensorineural hearing loss [6]. Among the audiometric techniques that can be used to assess the hearing are Pure Tone Audiometry, Impedance Audiometry and Brainstem Auditory Evoked Response [16]. Although, research has dwelt much on the epidemiology and clinico-pathological characteristics of sensorineural hearing loss in SCD [2–6], there is paucity of information on the overall patterns of ear diseases among children with SCD in Nigeria despite the huge burden of the disease in the country. This study describes the overall prevalence and patterns of ear diseases among Nigerian children with SCD. In addition comparisons were made between the burden of ear diseases in children with SCD and their non-SCD counterparts.

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2. Patients and methods

2.1. Study setting

This study was conducted at the Paediatric and Otorhinolaryngology clinics of the Ekiti State University Teaching Hospital in Nigeria between April 2016 and March 2017. The institution is a tertiary hospital serving the local communities in the state and also receives referrals from the neighbouring states.

2.2. Study design and data collection

This was a cross-sectional descriptive study of ear diseases manifestations in children receiving care for SCD and their non-SCD counterparts. Consecutive children with SCD confirmed by haemoglobin electrophoresis and high performance liquid chromatography (HPLC), Biorad, USA Variant II, using the Beta thalassaemia short program and their non-sickle cell disease counterparts aged from 5years with or without otologic conditions, attending the paediatric haematology, outpatients' and otolaryngology clinics were recruited and studied. A convenience sampling technique was used and all children aged 5–17 years attending the above-mentioned clinics constituted the sample frame. The exclusion criteria included: lack of consent by caregiver or refusal to participate, acute non otological illness, hearing loss of central nervous system origin and/or chronic medical or neurologic conditions. SCD patients who were using hydroxyurea, and participants who were too young to respond to Pure Tone Audiometric tone bursts were also excluded. The purpose of the study was explained to the parents/caregivers and participants before recruitment into the study. Written informed consents were obtained from the parents/caregivers as well as assents from participants aged 8 years and above. Only those whose parents gave consent to participate were recruited for the study. Those who met the study criteria were all seen and reviewed at both the paediatric and otorhinolaryngology clinics. Diagnoses of otologic diseases were made with detailed history, clinical examination, appropriate laboratory investigations and audiometric evaluation. Samples were taken from the ears of patients with suppurative otits media for microscopy, culture and sensitivity. The 'Triveni, TAM – 50' was used for the pure tone audiometry measurements while Otopront – Happersberger otopront GmbH was used for the impedance and acoustic reflex studies. Threshold levels in the ears were determined at 250, 500, 1000, 2000, 4000 and 8000 Hz. All were performed in a quiet room. The degree of hearing loss was classified according to the World Health Organisation grades, into: Mild (26–40 db), Moderate (41–60 db), Severe (61–80 db) and Profound (> 80 db), WHO Grading of deafness: (www.who.int/pbd/deafness/grades_of_hearing.PNG?ua=1). The average of at least, two steady state measurements of the Haemoglobin concentration (Hb conc) and Fetal haemoglobin (HbF) measured with automated Sysmex KX21N haematology analyser (Sysmex Corporation, Japan) and HPLC respectively were obtained from the records of the SCD participants. The Hb conc and HbF of the non SCD participants were taken on the day of recruitment or as soon as possible thereafter. For the SCD participants, steady state was defined as being free from any acute event(s) for at least one month and transfusion free for at least three months prior to recruitment.1 The primary outcome of the study was presence or absence of ear disease while the secondary outcomes were types of ear diseases found among participants and interventions received.

2.3. Ethical consideration

Institutional approval with Ethical Clearance protocol number EKSUTH/A67/2016/04/0010B was obtained from the institution's Ethics and Research Committee. Written informed consents were obtained from the parents/caregivers of the participants.

Table 1
Comparison of biodata and haematologic parameters among participants.

Parameters	Sickle cell disease N = 84 Median (Range)	No sickle cell disease N = 84 Median (Range)	P value
Sex			
Male	48	54	0.4297†
Female	36	30	
Age in years	9.0 (5.0–16.0)	9.0 (6.0–17.0)	0.239*
Fetal haemoglobin (HbF%)	9.3 (0.2–24.4)	0.6 (0.1–2.8)	< **0.0001***
Haemoglobin concentration (g/dL)	7.5 (6.4–9.7)	11.4 (8.0–14.1)	< **0.0001***

NB: Statistical significant p values are in bold fonts, †= Fisher's Exact Test, *=Mann-Whitney Test.

2.4. Data analysis

The GraphPad Prism Program, version 5 for Windows (San Diego, California, USA) was used for the statistical analysis. The normal distribution of the quantitative variables was verified by the Kolmogorov-Smirnov and Shapiro-Wilk tests. Continuous variables with non-normal distribution were expressed in median and analyzed by the Mann-Whitney tests for comparison of two independent groups. Chi-Square test or Fisher's Exact Test was used to compare categorical variables as applicable and level of statistical significance was set at $P < 0.05$.

3. Results

In all, there were 84 participants with SCD (81 Hemoglobin SS (SS) & 3 Hemoglobin SC (SC) and 84 without SCD, out of which 66 (39.3%) and 102 (60.7%)) were females and males respectively. There was no significant difference in both sexes and age of the two groups of participants. However, the Hb conc of the SCD group was significantly lower while their HbF level was significantly higher when compared with the non-SCD group. (Table 1).

Overall, the number of participants with any ear pathology was higher among the SCD participants relative to the control group 51 (60.7%) vs 29 (34.5%); p = 0.0011. Similarly, the number of ears with diseases were higher among the SCD compared to the non SCD group {80 (47.62%) versus 37 (22.02%)}; p < 0.0001 (Table 2). The most common ear disease among the SCD was SNHL and this was found among 26 (30.9%) of them – 23 isolated, 2 and 1 co-existing with otosclerosis and otitis media with effusion respectively. The most common ear pathology among the control group was wax and this was found among 21 (25.0%) of their ears. There was a significant difference in the prevalence of sensorineural hearing loss (SNHL) between the sickle cell patients and the controls (P < 0.0001). Also, the total number of participants' ears with solitary otosclerosis shows significant difference between the two groups (P = 0.029). (Table 2).

All the 3SC participants had SNHL. Majority of the sensorineural hearing loss were mild. Fig. 1.

All the five non-SCD patients with any sensorineural loss had mild forms of sensorineural hearing loss. Upon analysis for the influence of haematologic parameters on SNHL among the SCD patients, we found that there was no significant difference between the Hb concentration of SCD patients who had SNHL or did not have SNHL: 7.6 g/dl (6.6–9.4) vs 7.4 g/dl (6.4–9.7) respectively (P = 0.235). Fig. 2.

Similarly, the HbF levels of the SCD patients did not discriminate between SCD participants with or without SNHL: median 9.4% (0.9–20.6), vs median 9.1% (0.2–28.5) respectively, (P = 0.957). Fig. 3.

Of the 13 ears (10 of SCD cohorts and 3 of control group respectively) with acute and chronic suppurative otitis media, bacteria isolates from 8 (61.5%) of the ears were pseudomonas aeruginosa and this comprises of 6 of the 10 suppurating ears of the SCD participants and 2 of their non-SCD counterparts. Isolates from the remaining 5 (4 SCD