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Management challenges of congenital & early onset childhood hearing loss in a sub-Saharan African country

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ABSTRACT

Background: Hearing impairment is a hidden human disability with potentially catastrophic and age long consequences. This study highlighted the challenges associated with the management of congenital and early onset childhood hearing loss in a sub-Saharan African country.
Methods: A retrospective descriptive study of children seen between January 2008 and December 2013
Result: A total of 223 children consisting of 124 (55.6%) males with (M:F) of 1.3:1. Age ranged 1–15 years (mean ± SD; 6.39 ± 4.37 years) and age group 1–5 years constituted the largest proportion (56.5%). Congenital causes, febrile illness and hypoxia were the leading causes of HI. Over 93% had moderately severe to profound hearing loss and 64.6% had delayed speech development. Majority (99.3%) with congenital/perinatal onset of HL had significantly delayed speech development and 99.3% of HL due to ototoxicity and infective causes had peri/post lingual speech impairment. Larger percentage of patients presented late; 16.6% of patients with congenital/perinatal onset of HL presented within the first year, >41% presented after the fifth year. Less than 5% had hearing aid fitted and patients with profound hearing impairment were referred for cochlear implant
Conclusion: The burden of congenital and early onset hearing impairment is high and management outcomes are unsatisfactory in our locality, Challenges associated with managing such children were discussed, and suggestions/strategies for better management and outcome were made.

1. Introduction

Hearing impairment is a hidden human disability with potentially catastrophic and age long severe consequences [1]. Its consequences are comparatively more noticeable and more devastating in children. Compared with the developed countries, the burden, grave consequences and negative impact of hearing impairment (HI) is more in the developing countries of South Asia, Asia pacific and sub-Saharan Africa (SSA) where majority of the children with significant hearing loss are born [2–5]. In fact, childhood HI has become a major public health problem in sub-Saharan Africa (SSA). Notable among the consequences of hearing loss in children is the delay in development of language and speech manifesting as speech disorders [1,2]. Which negatively impact their lives. The effect of speech delays and disorders is multi-faceted and includes learning disorders, academic failures, communication and emotional – behavioral disorders and delay in social – cultural growth [2,6,7]. Without an intervention, this negative impact of hearing impairment continues into adulthood and its effects are varied in significance [2].

Management of the hearing impaired child involves an early identification, proper characterization and treatment, rehabilitation and possible prevention. In the United States and other developed countries, a lot of progress had been made in the management of hearing impaired children in the past decade; for example, passage of the Americans with Disabilities Act (ADA) has helped improve awareness, access, and healthcare for patients with hearing loss. Also, implementation of newborn hearing screening program coupled with provision of wide variety of social services and accessibility programs to those who are diagnosed with a hearing loss had assisted in rehabilitation of such children [5]. In Nigeria, (sub-Saharan Africa) there are many pressing health concerns to address and consequently HL, is given less priority despite the significant burden it poses. There is generally lack of information and awareness about HL and its consequences. Furthermore, many patients present late which further delays intervention strategy that has been demonstrated to effectively limit the negative impact of HL on the child's development [2,8].

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This study appraise cases of congenital and early onset hearing impairment that presented at the ENT clinic of a tertiary hospital in Nigeria, and aims to describe the types and patterns of HI, highlight major challenges confronting effective management of the condition, and proffer solution to mitigate the effects of childhood hearing loss in Nigeria, and by extension in other sub-Saharan African countries.

2. Methodology

This study was a retrospective descriptive study conducted between January 2008 and December 2013 at the ENT department of the LAUTECH Teaching Hospital, Osogbo, Nigeria. All patients diagnosed with congenital or early onset childhood HL were included in the study. Data were obtained from the clinical records of patients and information retrieved included socio-demographics, pregnancy history, delivery and immediate post-delivery events, history of febrile illness, time of onset of hearing loss, family history, and audiometric findings.

Pure tone audiometry (PTA) evaluations were done for children aged 5 years and above who had residual hearing and could respond appropriately to instructions, while the others were screened with free field audiometry and some had auditory brain stem response (ABR) audiometry. The degree of hearing loss for the patients was based on the World Health Organization (WHO) standard classification [3,4]. PTAs were performed using a diagnostic audiometer Elkon 3N3 Multi (Elkon, India), calibrated yearly to International Organization for Standardization (ISO) standards in a sound-proof room. Pure tone average (PTAv) values were calculated as the arithmetic mean of the air-conduction hearing thresholds at frequencies of 0.5, 1, 2, and 4 kHz. ABR was performed with MB II standard (classic) class IIa (MAICO Germany).

Patients' information including the audiological parameters was recorded. Data of patients with incomplete information and those that were lost to follow-up were excluded from the study. The data were entered into a spreadsheet and presented in a descriptive form as proportions, means and standard deviations as appropriate, in tabular and graphic forms. A statistical analysis was performed using statistical package for social sciences (SPSS) version 14 (Chicago, IL).

3. Results

Information of 223 children with HI were obtained for the study consisting of 124 (55.6%) males with (M:F) of 1.3:1. The age of the patients ranged from 1 to 15 years (mean ± SD = 6.39 ± 4.37 years). Majority (56.5%) of the patients were in the age group 1 to 5 years. Larger percentage of the patients presented late; 25 (16.6%) among patients with congenital/perinatal onset of hearing loss presented within the first year, 62 (41.3%) presented after the fifth year ($^{***}\chi^2 = 40.648$, $p = 0.001$). All the 151 patients with congenital/perinatal onset of hearing loss presented to primary care physicians (Paediatricians, Family physicians/general duty doctors) before referral to specialist clinic. Table 1 shows the age, gender and time of presentation distribution among the patients. Fig. 1 shows probable causes of hearing impairment among the children. Etiological diagnosis was based on history of event(s) that preceded onset of HI. Idiopathic/congenital hearing impairment (50.7%), hypoxia (13.9%) and measles/mumps (febrile illness) (13%) were the leading causes of HI in the patients. Preventable causes were responsible for 109 (48.9%) of the cases. Fig. 2 shows the degree of hearing impairment among the affected children in both ears. Majority of the children (94.6%) had educationally significant hearing loss at presentation while 64.6% had delayed speech development. Majority 149 (99.3%) of children with HI due to congenital causes, birth asphyxia and neonatal jaundice had significantly delayed speech development while 45 (99.3%) of those with HI due to ototoxicity and infective cause had peri/post lingual speech impairment ($\chi^2 = 2.243, p = 0.000$) Table 2. Table 3 determined the degree of association between speech impairment and age of onset, aetiology and threshold of hearing impairment using correlation coefficient. It revealed that age of onset and aetiologies of hearing impairment significantly correlated with speech impairment ($p < 0.05$).

Table 1
Age, gender and the time of presentation among the patients.

Variables	Number	Percentage	
Age in years			
≤1.0	20	9.0	
1.1–5.0	94	42.2	
5.1–10.0	57	25.6	
10.1–15.0	52	23.3	
Mean ± SD	6.39 ± 4.37		
Gender			
Male	124	55.6	
Female	99	44.4	
Time of presentation (among patients with congenital/perinatal hearing loss)			
Time of presentation	Number (%) ($N = 151$)	Chi square	p-Value
Within 1st year	25 (16.6)		
2nd year	29 (19.3)	40.648	0.001
2nd–5th year	34 (22.7)		
>5th year	63 (41.3)		

Forty six patients (20.6%) had enrolled at school for the deaf before presentation to Otolaryngologist. Less than 5% had hearing aid fitted followed by auditory and speech training with good outcome and patients with profound hearing impairment were referred for cochlear implantation.

4. Discussion

Management of a child with hearing impairment is one of the major and multi-faceted challenges confronting practicing Otorhinolaryngologists, pediatricians, social workers and geneticists in sub-Saharan African countries. The multi-faceted challenges aggravated by seeming lack of attention to early diagnosis, had made treatment and rehabilitation of hearing impaired children elusive in many developing countries. Findings from this study show that preventable causes are still prevalent in our environment constituting 48.9% of the causes of HI. Knowledge about preventable causes of hearing impairment has allowed prevention of large proportion of HL in developed countries, as reflected by its reduced incidence [2]. Conversely this knowledge has had little impact on prevention of HL in developing countries, where incidences continue to rise [5,9–12].

Measles is no longer a major cause of HL in developed and some developing countries consequent upon effective immunization programmes [2,9]. The few cases of hearing loss from measles in our study might be related to non-or ineffective immunization, resulting from break in the cold chain of the vaccines from irregular power (electricity) supply or ineffective vaccine. Moreover the National programme on immunization (NPI) in Nigeria at present excludes vaccinations for mumps, rubella and meningitis, which are notable risk factors for hearing loss [13] and as seen in this study. Although several effective vaccines against meningitis had been developed for use in the "meningitis belt" (which includes Nigeria) the vaccine is yet to be implemented into the NPI [2,13]. We advocate improvement upon the present level of childhood immunization programs to include vaccines that are viable and potent, and promote wider and effective coverage. Birth asphyxia and difficult delivery had been noted significant factors associated with increased risk of developing hearing loss in sub-Sahara Africa [14,15]. Birth asphyxia/hypoxia was a notable cause of HI in this