AETIOLOGY OF PROFOND BILATERAL SENSORINEURAL HEARING LOSS IN CHILDREN: CLINICAL SPECTRUM IN NORTH-EASTERN NIGERIA.

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ABSTRACT

BACKGROUND: profound bilateral sensorineural hearing loss has being a major hearing disability in children worldwide, most especially in tropical sub-Saharan Africa. Various aetiological factors ranging from congenital causes, maternal and childhood infections to the use of ototoxic drugs have been implicated. This study aims at highlighting the various aetiological factors and the pattern of profound bilateral sensorineural hearing loss in children.

METHODOLOGY: Case files of two thousand, seven hundred and sixty-one children aged 1-15yrs who presented to our facilities during the study period, with all types and grades of hearing loss were reviewed. Out of this four hundred and thirty-six (436) were children that presented with profound bilateral sensorineural hearing loss; the obtained data was documented on a structured pro forma questionnaire for statistical analysis. The variables include the socio-demographic data of the children, the various aetiological factors of hearing loss at presentation, laboratory investigations; and detailed clinical and audiological assessment.

RESULTS: Among the 2761 case files of children with both conductive and sensorineural hearing loss that was retrieved, 436(15.8%) with profound bilateral sensorineural hearing loss was analyzed. There were 264 males and 172 females, with M: F =3:2, age-group of 1-4yrs was found to be the most affected with 184.0(42.2%).Generally more males affected than females. There was a decrease in the frequency of profound sensorineural hearing loss with increasing age. Unknown (idiopathic) causes were found to be the commonest
aetiological factor with 144.0 (33.0%), this was followed by measles and meningitis infections with 21.6%, and 14.4% respectively.

CONCLUSION: Profound bilateral sensorineural hearing loss still remains a major childhood disability in our environment. More efforts should be made towards providing facilities for effective virology and genetic studies in our health care institutions. Intensive immunization campaign, especially against measles and meningitis should be emphasized nationally.

KEY WORDS: PTA (pure tone audiometry), Measles, sensorineural hearing loss (SNHL), children.
INTRODUCTION

Profound bilateral sensorineural hearing loss in children still constitutes a major source of disability worldwide. United Nations statistics, estimated that there are 350 million children in the developing world whose facilities are below the minimum standard, out of these already disadvantaged children, 5 million are profoundly deaf\(^1\). In sub-Saharan Africa; infections still play a major role in the aetiology. The commonest infections include Measles, mumps, rubella and meningitis. Studies conducted in Nigeria, and in some West African countries showed that Measles and meningitis are still the commonest infections associated with profound sensorineural hearing loss\(^2,3,4\).

The prevalence of measles as an infective aetiological factor has been found to be 13.5% and 19.3% in Nigeria\(^2,5\), while in some West African countries it has been found to be 45.0% (Sierra Leone),\(^2\) and 30.0% (Ghana)\(^5\).

Nigeria being part of the meningitis belt of West Africa, the prevalence of meningitis-induced sensorineural hearing loss vary among regions. The far-north which is dry, arid with frequently dusty winds has a higher prevalence of 21.3%\(^6\), when compared to the southern part of the country with a prevalence of 8.7%\(^5\). The implications of profound sensorineural hearing loss in children will be more manifest in the north-east, given the geographical location and the educational background of the region.

This study aims at highlighting the various aetiological factors and the pattern of profound bilateral sensorineural hearing loss in children, in north-eastern Nig
METHODOLOGY

The university of Maiduguri teaching hospital is among the largest tertiary health institutions in the North-East. The ear, nose and throat department of the hospital receives referrals from within the hospital, from the five north-eastern states and also from the neighbouring countries of Chad, Niger, and Cameroon republics.

This was a hospital based retrospective study over a four-year period (January 2007 to December 2010). The study involved all children aged 1-15 years, who presented to the ear, nose and throat clinic and the in-patient section of the department with hearing disability.

A questionnaire pro forma was designed for data collection, the data extracted include, the child’s demographic data, which includes age, gender, case file hospital number and residential address. Others were the onset and duration of hearing loss, history of hearing loss in the family, history of ototoxic drugs (common drugs cited).

All the patients had full clinical evaluation including pure tone audiometric evaluation. Ear, Nose and throat examination was carried out in the ENT clinic in accordance with standard procedure. Patients with soft cerumen auris had manual removal with Jobson Hornes probe or syringing with warm normal saline at body temperature. Those with impacted cerumen auris had them softened using waxol or olive oil for three or four days before syringing. Patient’s hearing acuity was initially assessed crudely with the use of a turning fork 512 Hz for Rinne’s and weber’s test only or a distraction test in children <5yrs old. Patients were then referred to the audiology room where an audiological assessment was conducted. Children less than 5yrs of age had free field audiometry and their response to
various sounds was documented. Older children above the age of five-years had pure tone audiometry. (Audiometer type DA 931 mains operated with a voltage stabilizer equipped with well-fitting standard head phones). The audiometer was calibrated every six months in accordance with the world health organization ear and hearing disorders survey protocol \(^7\). Testing was carried out in a double walled sound proof booth in accordance with standard procedure \(^8\).

Degree of hearing loss was assessed using pure tone average of air conduction thresholds at 0, 5, 1, and 2KHZ for each ear; these were then categorized in accordance with the, World Health Organization’s grading of hearing impairment, \(^9\) as follows:

<table>
<thead>
<tr>
<th>Degree of Hearing Loss</th>
<th>DB Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Within normal limits</td>
<td>&lt;25db</td>
</tr>
<tr>
<td>Mild hearing loss</td>
<td>26-40db</td>
</tr>
<tr>
<td>Moderate hearing loss</td>
<td>41-60db</td>
</tr>
<tr>
<td>Severe hearing loss</td>
<td>61-80db</td>
</tr>
<tr>
<td>Profound hearing loss</td>
<td>&gt; 81db</td>
</tr>
</tbody>
</table>

Hearing loss was considered conductive when the air-bone gap was equal to or greater than 15db.Data obtained was analyzed using the statistical package for the social sciences (SPSS) software, version 16.0
RESULTS

Case records of 2761 children aged 1-15 yrs who presented to our facilities with all types and grades of hearing loss during the study period were retrieved from the medical health records and reviewed. 436(15.8%) were found to have profound bilateral sensorineural hearing loss.

There were 264 males and 172 females, with a male to female ratio of 3:2. The age and gender distribution of the patients showed a male preponderance in all the age groups, except in the age group 5-9 yrs which showed a female preponderance with 19.5%. The various aetiological factors found are as in table-ii, with the unknown constituting the highest percentage, 33.0% (144)

Pure-tone audiometry and free-field audiometry done on the patients showed that out of the 2761 cases that presented with all forms and grades of hearing loss, 436(15.8%) were specifically found to have profound bilateral sensorineural hearing loss.
Table-i: Age and Gender distribution of the children

<table>
<thead>
<tr>
<th>Age (yrs)</th>
<th>Male (%)</th>
<th>Female (%)</th>
<th>Total (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1-4</td>
<td>154 (35.3)</td>
<td>30 (6.9)</td>
<td>184 (42.2)</td>
</tr>
<tr>
<td>5-9</td>
<td>36 (8.3)</td>
<td>85 (19.5)</td>
<td>121 (27.8)</td>
</tr>
<tr>
<td>10-14</td>
<td>49 (11.2)</td>
<td>40 (9.2)</td>
<td>89 (20.4)</td>
</tr>
<tr>
<td>&lt;15</td>
<td>25 (5.7)</td>
<td>17 (3.9)</td>
<td>42 (9.6)</td>
</tr>
<tr>
<td>Total</td>
<td>264 (60.5)</td>
<td>172 (39.5)</td>
<td>436 (100.0)</td>
</tr>
</tbody>
</table>
### Aetiology of profound bilateral sensorineural hearing loss

<table>
<thead>
<tr>
<th>Causes</th>
<th>No. (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Unknown</td>
<td>144.0 (33.0)</td>
</tr>
<tr>
<td>Measles</td>
<td>94.0 (21.6)</td>
</tr>
<tr>
<td>Meningitis</td>
<td>63.0 (14.4)</td>
</tr>
<tr>
<td>Febrile Illness</td>
<td>38.0 (8.7)</td>
</tr>
<tr>
<td>Trauma</td>
<td>32.0 (7.3)</td>
</tr>
<tr>
<td>Ototoxicity</td>
<td>22.0 (5.0)</td>
</tr>
<tr>
<td>Severe Birth asphyxia</td>
<td>17.0 (3.9)</td>
</tr>
<tr>
<td>Mumps</td>
<td>11.0 (2.5)</td>
</tr>
<tr>
<td>Neonatal jaundice</td>
<td>8.0 (2.0)</td>
</tr>
<tr>
<td>Cerebral palsy</td>
<td>7.0 (1.6)</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>436.0 (100.0)</strong></td>
</tr>
</tbody>
</table>
DISCUSSION

Profound bilateral SNHL has been a major source of disability in children. In this study there were 264 males and 172 females with M: F =3:2, this is similar to earlier studies in a West African sub-region\textsuperscript{4}. Among the aetiological factors, infections still play a leading roll; commonest among the infections are meningitis, measles and mumps.

Analysis of these showed that age group 1-4yrs had 184.0(42.2%) patients, age group 5-9 with 121.0(27.8%), age group 10-14yrs 89.0(20.4%), while age group less than 15yrs had 42.0(9.6%) patients. This showed a decline in the frequency of hearing loss with increasing age which is in agreement with earlier studies\textsuperscript{2} this could probably be due to early detection of hearing disability by the parents which necessitates early health-seeking behavior. 436.0(15.8%) presented with profound bilateral SNHL. This is in contrast to the findings of Ahmad in Ibadan (Nigeria) with 44.9\textsuperscript{10}. This could be due to a difference in sample size, even though in both studies profound SNHL was found to be leading.

Idiopathic (unknown) cause was found to be the commonest, this is similar to that found in some earlier studies conducted in Nigeria\textsuperscript{2}, but is dissimilar with some studies done in some west African countries\textsuperscript{3,4}. The lack of facilities for genetic studies could be the reason for this high prevalence, because genetic factors account for at least 80% of congenital deafness, largely due to mutations in connexin-26 gene\textsuperscript{11}. And in 90% of cases the children were born from parents with normal hearing\textsuperscript{12}. Measles infection constitutes 94.0(21.6%); this is in contrast to some earlier studies done in Nigeria\textsuperscript{2,5}. The hallmark of measles prevention is immunization; the global immunization coverage has reached an
estimated 80% in 2006\textsuperscript{13}. The perception that vaccines contain some unsafe agents, could be the socio-cultural factors mitigating against effective immunization coverage in the north-east, hence, some measure of rejection cannot be ruled-out as has happened to polio-vaccine in the past\textsuperscript{14}. Effective coverage is attained with a second-dose schedule which is advocated because it reliably leaves 99.0% of those vaccinated immune\textsuperscript{15}. Meningitis survivors constitutes 63.0(14.40%), this is in contrast to the earlier studies\textsuperscript{2,3,4}. This could be due to the study area ,which is dry, arid savannah, lies in the meningitis-belt of sub-Saharan Africa\textsuperscript{16}. Survivors, in 30% of cases, are known to sustain at least 30db SNHL after acute bacterial meningitis\textsuperscript{17}. Mumps infection constitutes 11.0(2.5%); this is in contrast to some earlier studies\textsuperscript{2}. The low prevalence could be due to late detection or due to undetected cases, since mumps virus is known to cause unilateral SNHL which makes early detection difficult in children\textsuperscript{3}.

Febrile illness with or without convulsions constitutes 8.7%; this is similar to that found earlier in Nigeria\textsuperscript{18}. This could be due to the frequent use of anti-malarial drugs and antipyretics as OTC (over-the-counter) drugs which possibly alters the health-seeking behavior of parents to professionals where early diagnosis should have been made. Head and neck injury constitutes 7.3%, this in contrast to earlier studies\textsuperscript{3,19}. The increase in the prevalence could be due to the numerous commercial motorcycles in recent times, associated with a rise in road traffic accidents. Ototoxicity constitutes 5.0% in contrast to that found by ologe et al\textsuperscript{2}. This could be due to the indiscriminate use of ototoxic drugs\textsuperscript{20}. Mitochondrial A1555G gene mutation which causes gene carriers to be exquisitely sensitive to the ototoxic effects of these drugs is the most common cause of genetic deafness in countries where these antibiotics were indiscriminately abused\textsuperscript{21}.
Our study showed a low prevalence of perinatal conditions associated with profound bilateral sensorineural hearing loss, such conditions include, severe birth asphyxia, 3.9%, neonatal jaundice, 2.0%, and cerebral palsy 1.6%. This is similar to that found in earlier studies, but in contrast to some studies in Nigeria.

This could partly be due to the improved perinatal care in our institution, and partly due to the larger majority of perinatal care being provided by the traditional birth attendants (TBA) at home, which may have reduced hospital attendance.
CONCLUSION

We thus conclude that causes of profound bilateral sensorineural hearing loss of unknown aetiology is still of paramount significance in our environment, this is in addition to the common infective causes.

Health care institutions should be properly equipped with modern diagnostic facilities, so as to enhance effective diagnosis of viral and congenital causes of sensorineural hearing loss. Congenital hereditary causes can be prevented by discouraging consanguineous marriages.

Resources for genetic studies should be provided and supported in all our health care institutions so as to prevent or enhance early diagnosis of congenital causes of profound sensorineural hearing loss. Encourage immunization against the infective diseases.
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