A RETROSPECTIVE STUDY OF THE HEARING LOSS IN MICROCEPHALY CHILDREN
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ABSTRACT

BACKGROUND
Microcephaly is defined as a head circumference more than two standard deviations below the mean for gender and age. Congenital microcephaly is present at birth, whereas postnatal microcephaly occurs later in life and associated with developmental delay in children. Genetic abnormalities, syndromes, metabolic disorders, teratogens, infections, prenatal, perinatal, and postnatal injuries can cause both congenital and postnatal microcephaly. In patients with microcephaly and global developmental delay, hearing loss may affect further development. We wanted to assess and analyse the audiological profile in children with microcephaly.

METHODS
A cross sectional retrospective and analytical study was conducted by studying the charts and medical records of 121 children with developmental delay for a period of six month with permission of ethical committee of the institution. The children included in this study were those who came to paediatric ENT OPD of Christian Medical College, Vellore. Out of 121 children studied, 44 children had microcephaly. Hospital records from medical records section were taken and history of consanguineous marriage, detailed paediatric birth history and developmental milestone records were noted. All children had undergone detailed ENT examination including microscopic examination of the ear. Audiological evaluation was done by Behavioural Observation Audiometry (BOA), Oto Acoustic Emission (OAE) and Brainstem Evoked Response Audiometry (BERA), Tympanometry (Tymps). They showed hearing loss which was analysed. Degree of hearing loss was classified using American Speech-Language Hearing Association ASHA classification.

RESULTS
Among 44 children with microcephaly, there were 27 (61.4%) males. The mean age of the study group was 3.1±1.05 years. The age group below 6 yrs. in this cohort was 36 (81.8%). The youngest child in the study was 6 months old and oldest child being 10 years old. Behavioural observation Audiometry (BOA) results showed that out of 44 children, 30 (68.2%) with normal hearing and 14 (31.2%) had hearing loss. Consanguinity of marriage was seen in 7(15.9%). On examination of birth history records, these microcephaly children were born full term babies; 39 (88.6%) and 28 (63.6%) were appropriate to age. 21 (47.7%) children cried immediately at birth. Chi square test was applied, and the results show that microcephaly was not associated with hearing loss.

CONCLUSIONS
Among the 44 microcephaly children included in the study, 14 (31.2%) had hearing loss. Our study detected a higher incidence of undetected hearing loss among 14 (31.2%) children with microcephaly. It was concluded that children with microcephaly should be routinely screened for potential hearing loss. On statistical analysis no association of hearing loss with microcephaly was observed.

KEYWORDS
Hearing Loss, Microcephaly, BERA, Consanguinity

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abnormal foetal development. Microcephaly has been associated with intellectual disability and developmental delay. In addition to congenital microcephaly, there is also an acquired form of microcephaly in which an infant’s head circumference falls within the normal range at birth with subsequent development of microcephaly over time due to deceleration of brain growth. Children with global developmental delay and microcephaly require complex, individualized therapy to maximize their long-term quality of life. However, the effects of hearing impairment are amenable to the treatment and rehabilitation strategies if identified at an early age and effective intervention program can be initiated. Thus, overall future and success of a child can be improved by reducing the complications of hidden disability of hearing impairment. Metabolic disorders are more likely to cause postnatal onset microcephaly and are typically associated with global developmental delay (GDD).³ However, there is no data is available in the Indian context, with regards to the degree, and type of hearing impairment in children with global developmental delay and microcephaly. Hence no retrospective study was done to find the presence of hearing impairment in children with microcephaly and global developmental delay. Our aim is to increase the awareness of possible correctable audiological impairment that hinder development and learning in children with microcephaly with global developmental delay children.

We wanted to describe the clinical and audiological profile in children with microcephaly and developmental delay presenting to Pediatric ENT unit and determine the type and severity of hearing impairment in these children with microcephaly.

**METHODS**

This retrospective study was done by studying the charts and medical records of children with microcephaly and Global developmental delay who presented to the ENT OPD for six month.

**Inclusion criteria**

All children with microcephaly and global developmental delay from six months to sixteen years age attending ENT-OPD were included.

**Exclusion Criteria**

Children aged below 6 months and above 16 years, children with mental retardation and epilepsy were excluded and children on sedatives and anti-convulsion therapy were excluded.

### Degree of Hearing Loss

<table>
<thead>
<tr>
<th>Degree of Hearing Loss</th>
<th>Hearing Loss Range (dB HL)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>-10 to 15</td>
</tr>
<tr>
<td>Minimal</td>
<td>16 to 25</td>
</tr>
<tr>
<td>Mild</td>
<td>26 to 40</td>
</tr>
<tr>
<td>Moderate</td>
<td>41 to 55</td>
</tr>
<tr>
<td>Moderately severe</td>
<td>56 to 70</td>
</tr>
<tr>
<td>Severe</td>
<td>71 to 90</td>
</tr>
<tr>
<td>Profound</td>
<td>91+</td>
</tr>
</tbody>
</table>

*Table 1. American Speech-Language Hearing Association (ASHA) Classification*

From the medical records and charts detailed ENT examination were noted and from the audiological records of this patient studied which included Behavioural observation audiometry (BOA), Oto Acoustic Emission (OAE) and Brainstem Evoked Response Audiometry (BERA), Tympanometry (Tymps). Brainstem evoked response audiometry was done using Intelligent hearing screening machine to assess hearing loss. In BERA, click stimulus of 60 to 90 dB above hearing threshold in the frequency 2 KHz to 4 KHz and tone burst both are used. The stimulus rate used was 30.1 clicks/sec. Active electrode (red) was placed over forehead, reference electrode (black) over ipsilateral mastoid or ear lobe and ground electrode over contralateral mastoid were placed. Tympanometry was done in all children to using Grason Stadler 61 Tympanometer. Degree of hearing loss was classified using American Speech-Language Hearing Association ASHA classification (Table 1).

Otoacoustic emission –Distortion product OAE reports were recorded and Tympanometry findings were noted down. Appropriate statistical tests were used to analyse the data. All analyses were done using Statistical Package for Social Services (SPSS) software Version 21.0. The Chi-square or Fisher’s exact test was applied to the data using microcephaly with developmental delay and non-Microcephaly children with developmental delay.

**RESULTS**

Medical records and charts of One hundred and twenty-one children with developmental delay were studied retrospectively. All the children detailed ENT examination including examination under microscope of ear and audiological assessment were studied with the charts from ENT department and Developmental paediatric department.

Out of 121 charts reviewed 44 had microcephaly with developmental delay. Majority of the subjects were males; 27 (61.4%). The mean age of the study group was 3.1±1.05 years. The age group below 6 yrs. was 36(81.8%). The youngest child in the study was 6 months old and oldest child being 10 years old. Consanguinity of marriage between the parents was studied. In the present study non-consanguineous marriages were 37(84.1%) and consanguineous marriages between parents were 7(15.9%). On birth history record the microcephaly children were born full term baby 39(88.6%) and 28 (63.6%) were with normal birth weight and 21(47.7%) cried immediately at birth. Out of 44 microcephaly children behavioural observation audiometry result showed 30 (68.2%) had normal hearing and 14 (31.8%) had hearing loss. Otoacoustic Emissions findings in this study showed Out of 44 children right ear OAE were absent in 19(43%) ears and present in 9(20.5%) ears. Left ear OAE result show absent in 16(36.4%) and present in 12(27.3%). The findings on Brain Evoked Response Audiometry (BERA) showed out of 44 children examined for BERA on right ear 15(34.1%) had normal hearing, 2(4.5%) had minimal loss, 4(9.1%) had mild hearing loss, moderate hearing loss in 8(18.2%) ears, moderately severe in 2(4.5%), severe in 2(4.5%) ears, profound in 7(15.9%) ears and not done in 4(9.1%) ears.
BERA on left ear 15(34.1%) had normal hearing, 1(2.3%) had minimal loss, 6(13.6%) had mild hearing loss, moderate hearing loss in 8(18.2%) ears, severe in 3(6.8%), profound in 7(15.9%) ears and not done in 4(9.1%) ears. Findings of Tympanometry were Out of 44 children with microcephaly on right ear A type curve, 24(54.5%) B type curve in 8(18.2%) and 7(15.9%) have C type curve. Out of 44 children with microcephaly on left ear A type curve, 24(54.5%) B type curve in 9(20.5%) and 6(13.6%) have C type curve. Associations between sensorineural hearing loss and microcephaly were analysed and testing using Fisher's exact test, in these one hundred and twenty-one children with control group and 44 with microcephaly, it was not significant statistically.

DISCUSSION
This retrospective observational study was done in tertiary care center for six months to find the clinical and audiological profile of children with microcephaly and global developmental delay by studying the records of the charts of these patients who had been seen in Otorhinolaryngology OPD after obtaining clearance from ethical committee of the institute. Children were recruited in this study were according the inclusion and exclusion criteria of the study. Various studies have attempted to understand the association between microcephaly and delayed development with hearing loss and mainly the data comes from developed countries. The paucity of studies on hearing assessment in microcephaly and global developmental delay children we did this retrospective study of children with microcephaly and global developmental delay and studied the audiological investigation such as BERA, Tympanometry and OAE from the medical records. This study addressed the hearing in children diagnosed with microcephaly and global developmental delay. The aim was to determine the pattern and frequency of hearing loss in a cohort of children with confirmed diagnosis of microcephaly and global developmental delay done by developmental paediatrician. Children with microcephaly and global developmental delay require complex, individualized therapy to maximize their long-term quality of life. If hearing loss is undetected early in developmental stage, it affects the speech and language and adds on to delay. Hearing loss being a potential cause for speech delay is often overlooked in these groups of children. Hence it is very essential to educate the paediatrician looking after the child that hearing assessment is a vital tool in the diagnostic armament of evaluating the child especially with suspected delay in development and microcephaly. The latest consensus definition used by the American Academy of Neurology (AAN) practice parameter statement defines global developmental delay operationally, as a significant delay in two or more developmental domains.4 In this study it was found that all the children with microcephaly and global developmental delay who were referred from developmental paediatrics with suspected to have hearing loss, the result in our study showed 15.9% had sensorineural hearing loss. There was a male preponderance in the study group with 27 (61.4%) males and 17 (38.6%) females. In our study the children were grouped into two categories according to the age, first group of children less than six years and the second group of children of six years of age or more. Majority 81.8% of the study population was in the under six age group. This can be attributed to the obvious delay in milestones of the child which compels the parents or care givers to seek medical help early. The mean age of the study population was 3 years. The youngest child in the study was six months and oldest child 10 years. However, this is not a standard practice of referral to Otornhinalaryngologist for microcephaly and global developmental delay at first visit, as usual practice is to identify hearing status by startle reflex and clapping test. In this study children with microcephaly and global developmental delay had their hearing assessment by BOA, OAE, BERA and tympanometry. Out of 44 children BOA shows 30(68.2%) had normal hearing and 14(31.8%) had hearing loss. Olusanya BO etal study showed increase risk of hearing loss in microcephaly children.5 Otoacoustic emissions were absent in 19(43.2%) in right ears, present in 9(20.5%) children and on left ear OAE was absent in 16(36.4%). Study done by Anju et al showed 16.9% hearing loss in microcephaly children with delayed development.6 Our study shows similar results, on BERA findings from medical records and audiological records showed that in these 44 children sensorineural hearing loss 15.9% in bilateral ear and 15 (34.1%) had normal hearing bilaterally.

CONCLUSIONS
Among children with microcephaly and global developmental delay who were included in the study, 15 (34.1%) had normal hearing and sensorineural hearing loss was present in 15.9% bilaterally. Our study detected a higher incidence of hearing loss by BERA testing 65.9% in children with microcephaly and global developmental delay. Our study recommends that every child of microcephaly with global developmental delay should have a detailed audiological work up to identify hearing loss and follow up for early identification of middle ear pathology so that timely intervention can be taken.

Limitations
The major limitation of this retrospective observational study was that it looked at a point estimate of hearing in children with microcephaly and global developmental delay who attended Otorhinolaryngology OPD. A second limitation of the study is that it is a hospital-based study and not a community-based study excluding children whose caregivers who didn’t seek medical advice for their children.

REFERENCES

