A STUDY OF OPHTHALMIC ASSOCIATION IN CHILDREN WITH SEVERE SENSORINEURAL DEAFNESS
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ABSTRACT

BACKGROUND
Vision plays a significant role in acquisition of skills such as sign language in children with hearing impairment. Auditory training after cochlear implant surgery is given in combination with visual gestures. Thus even subnormal vision will adversely affect their social development. Hence there is a need to do ophthalmic evaluation in children with severe and profound hearing loss.

In this study children with severe sensorineural deafness prepared for cochlear implant surgery were evaluated for the presence of ophthalmic manifestation.

MATERIALS AND METHODS
This is a retrospective hospital-based study from April 2017 to March 2018. Children under the age of 10 years with severe sensorineural hearing loss who were referred to us for ophthalmological evaluation were included in the study. Complete ophthalmic evaluation including anterior segment examination with slitlamp, detailed fundus examination and cycloplegic refraction were done. Syndromic associations were also studied.

RESULTS
Fourteen children were included in the study period. All the children showed severe sensorineural hearing loss in otoacoustic emission and brainstem evoked response audiometry tests. Maximum number of patients (9 patients) were below 3 years of age (64.7%). There was a female predominance (57.1%). 35.7% of our study population had ocular involvement. Hypermetropia was noticed in two children (14.2%). One patient (7.14%) had salt and pepper appearance of fundus along with other features of congenital rubella syndrome. Waardenburg syndrome with heterochromia iridis in one patient (7.14%) and Usher Syndrome (7.14%) with retinitis pigmentosa in another patient were also noted. Three patients (21.4%) had associated cardiac involvement.

CONCLUSION
The results of our study indicate that there is a high prevalence of ocular problems in children with severe sensorineural deafness. Coincidence of visual disorders with hearing impairment especially in the early years can negatively impact the development of child. Hence addressing the visual problems helps a lot for them to improve their communication skills.

KEYWORDS
Sensorineural Hearing Loss, Rubella Retinopathy, Hypermetropia, Retinitis Pigmentosa, Waardenburg Syndrome.


BACKGROUND
The prevalence of ocular abnormalities among the hearing-impaired children is higher than the general population of comparable age group.1 The high rate of ocular pathology in deaf patients is related to the fact that the retina and cochlea have the same embryonic origin during the sixth and seventh weeks of embryonic development. Hence prenatal insult and toxins causing congenital anomalies can affect both of them.

Hearing impaired children are heavily reliant on visual clues in order to develop efficient communication skills.2 Thus even a mild visual impairment may reduce the visual clues available to the child impeding their social and mental development.3 Thus early identification of dual sensory deficits allows early intervention and maximization of quality sensory input critical for development.3 Ophthalmologic evaluation in children with sensorineural hearing loss serves 2 purposes. The first
purpose is to identify the ocular abnormalities requiring early intervention. The second purpose is to find out the presence of associated hereditary syndromes involving both eye and ear. This will aid in the identification of other syndrome-associated abnormalities and will have implications for genetic counselling.3

**Aim of the Study**

This study was conducted to assess the prevalence of ophthalmic manifestation in children with severe sensorineural hearing loss.

**MATERIALS AND METHODS**

Cases of severe sensorineural hearing loss evaluated for cochlear implant surgery in ENT department of a tertiary hospital during one year period from April 2017 to March 2018 were included in this study. This was a retrospective case study. Ethical committee approval was obtained.

Relevant ocular history and family history were taken. History suggestive of congenital heart disease and consanguineous marriage were also elicited. Complete ocular examination such as assessment of best corrected visual acuity, anterior segment evaluation with slit lamp, posterior segment examination with 90D lens, direct and indirect ophthalmoscope with 20D lens were done. Cycloplegic refraction was done. Objective refraction was done by performing retinoscopy. Intraocular pressure was measured by Schiotz and applanation tonometer.

**Inclusion Criteria**

Children below the age of 10 years were included in this study. Children with severe sensorineural hearing loss of both sexes were taken into this study.

**Exclusion Criteria**

Children with conductive deafness were excluded.

**RESULTS**

In this study 14 patients clinically diagnosed as having severe sensorineural hearing loss in otoacoustic emission & brainstem evoked response audiometry tests were subjected to thorough ocular examination before cochlear implant procedure. More number of children (64.2%) presented below 3 years of age in our study. Likewise, female children (57.1%) were more in number compared to male children. The demographic distribution is given in table 1. Two out of fourteen patients (14.2%) had positive family history of severe sensorineural hearing loss in the mother. Parents of two children (14.2%) had consanguineous marriage (Table 1). 3 out of 14 patients (21.4%) had cardiac anomalies. One child had undergone surgery for Patent ductus arteriosus at her 3 years of age. Two children (14.2%) had associated developmental delay. The details of associated systemic anomalies are depicted in table 2.

Around 35.7% of children in our study showed ocular involvement. The distribution of ophthalmic manifestations are represented in pie chart 1. Two children (14.2%) were found to have hypermetropia on cycloplegic refraction.

Fundus examination of one child (7.14%) revealed salt and pepper appearance in posterior pole. Retinitis pigmentosa was noted in another patient (7.14%) with severe sensorineural deafness and delayed milestones. All these features help us in arriving at a diagnosis of Usher syndrome in that child. The presence of heterochromia iridis and sensorineural deafness were suggestive of Waardenburg syndrome in one of the children (7.14%). Thus, we noticed syndromic association in two patients (14.2%) (Table 3). After thorough ophthalmological evaluation 2 out of 14 patients underwent cochlear implant surgery. They are doing well with auditory evaluation. They have been advised to come for ophthalmic assessment once in 6 months.

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Number</th>
<th>Percentage</th>
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<tbody>
<tr>
<td>Sex</td>
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<tr>
<td>Male</td>
<td>6</td>
<td>42.9%</td>
</tr>
<tr>
<td>Female</td>
<td>8</td>
<td>52.1%</td>
</tr>
<tr>
<td>Age</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;3 years of age</td>
<td>9</td>
<td>64.2%</td>
</tr>
<tr>
<td>&gt;3 years of age</td>
<td>5</td>
<td>34.8%</td>
</tr>
<tr>
<td>Family History</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mother with sensorineural deafness</td>
<td>2</td>
<td>14.2%</td>
</tr>
<tr>
<td>Consanguineous marriage between parents</td>
<td>2</td>
<td>14.2%</td>
</tr>
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</table>

**Table 1. Demographic Details and Positive Family History**

<table>
<thead>
<tr>
<th>Systemic Association</th>
<th>Number</th>
<th>Percentage</th>
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<tr>
<td>1. Cardiac anomalies</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Patent Ductus arteriosus</td>
<td>2</td>
<td>14.2%</td>
</tr>
<tr>
<td>• Mitral and pulmonary Valvular stenosis</td>
<td>1</td>
<td>7.14%</td>
</tr>
<tr>
<td>2. Developmental Delay</td>
<td>2</td>
<td>14.2%</td>
</tr>
</tbody>
</table>

**Table 2. Presence of Systemic Anomalies**

**Pie Chart 1. Distribution of Ophthalmic Manifestation**

![Pie Chart 1. Distribution of Ophthalmic Manifestation](image-url)
Syndromic Presentation

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Number</th>
<th>Percentage</th>
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<tr>
<td>Present</td>
<td>2</td>
<td>14.3%</td>
</tr>
<tr>
<td>(one case of Usher syndrome with retinitis pigmentosa and another case with Waardenburg syndrome with heterochromia iridis)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Absent</td>
<td>12</td>
<td>85.7%</td>
</tr>
</tbody>
</table>

Table 3. Percentage of Patients with Syndrome Involving both Eye and Ear

DISCUSSION

Visual and auditory senses are responsible for more than 90% of information acquisition from the environment. Hearing impaired children are dependent upon vision for communication and learning. Hence, they should be given every opportunity to maximise their visual potential at the earliest age as possible. Similarly, cochlear implantation at an early age can help in regaining near normal speech and language development. Maximum number (64.2%) of children in our study group are less than 3 years of age. This may be because of increased awareness among the parents to notice and bring their children with severe hearing loss for getting them treated. In our study population female children were more compared to male children.

Approximately 50% of all cases of congenital deafness are genetic in origin. Hereditary hearing loss may be inherited by autosomal dominant, autosomal recessive, X-linked or mitochondrial inheritance. It might be reasonable for us to suspect a genetic cause for the child’s deafness if it runs in the family.

Detailed family history was taken. In our study mother of two patients (14.2%) had sensorineural deafness since childhood. Parents of two patients (14.2%) had consanguineous marriage. Both these observations were supported by the research article on Study of association of family history and consanguinity with congenital hearing loss.

Both the eye and inner ear are neuroectodermal derivatives which lie close to each other during intrauterine development. Hence the incidence of ophthalmic problems is quite high in patients with congenital and infantile severe sensorineural deafness.

Refractive errors are the most common ocular problems in hearing impaired children. Among the refractive errors hyperopia and astigmatism are more common. In our study also there exists a high prevalence of hypermetropia (21.42%). This is consistent with the results of earlier studies conducted on eye problem in children with hearing impairment which state that there is an immaturity in the axial length development in patients with congenital sensorineural deafness. Children found to have hypermetropia in our study were prescribed spectacles. Parents were advised to adhere with constant wearing of glasses. Hyperopia especially in the presence of hearing impairment may hinder the use of sign language and facial expression to communicate. Uncorrected refractive error and strabismus may lead to amblyopia.

Systemic work up of all these patients showed the presence of cardiac anomalies in 3 (21.4%) patients. Mitral and pulmonary valvular stenosis were noted in one (7.14%) patient. Of the two patients (14.9%) who had patent ductus arteriosus one girl child was found to have salt and pepper appearance of the posterior pole on fundus examination. She also had developmental delay with subnormal Intelligent Quotient. All these features were suggestive of congenital rubella syndrome with classical triad. This is in accordance with results from another study conducted on ocular manifestations of congenital rubella syndrome in a developing country. Children with rubella retinopathy may develop choroidal neovascular membrane at a later date. They are also more prone to develop diabetes mellitus with diabetic retinopathy and thyroid disorder in later life. Hence we have advised the parents to bring their child for regular follow up. Since the introduction of rubella vaccination, the incidence of congenital rubella syndrome is rare now a days. Congenital TORCH infection may also be associated with other manifestations like microphthalmia, cataract, glaucoma, chorioretinitis and pigmented retinopathy.

Syndromic hearing impairment may account for up to 30% of prelingual deafness. Various syndromes are associated with both ocular manifestation and congenital sensorineural deafness. In our study we found out one case of Waardenburg syndrome (7.14%) who presented with heterochromia iridis and sensorineural deafness. These clinical features are in accordance with a previous study conducted on Waardenburg syndrome by Carol L. Shields et al. We have come across one more syndromic association in which the patient had retinitis pigmentosa and delayed mile stones. All these features were suggestive of Usher syndrome.
Thus ophthalmic evaluation also aids in arriving the etiological diagnosis. In most of the studies on congenital sensorineural deafness refractive error was the commonest association. But we found out that fundus pathologies are also equally prevalent. Our study differs from other previous studies in that apart from treating refractive error our fundus findings also help in confirming the diagnosis of congenital rubella syndrome and Usher syndrome. Hence, we would recommend that all children with severe to profound hearing loss should undergo ophthalmological assessment soon after the diagnosis of sensorineural deafness.

CONCLUSION
Our study was done in a centre for cochlear implant surgery to assess the ophthalmic association. Early detection and treatment of visual problems in these children with severe sensorineural deafness will help in improving their language, social and communication skills. Even though the study population is less around 35.7% of children had ophthalmic manifestation. This shows a significant correlation between both of them. Since these hearing-impaired children use their sight to compensate for deafness early detection and treatment of ocular problems is of paramount importance.

REFERENCES